Solutions for Nasal Defects

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Abstract: Many studies on defects caused by trauma, infection, cancer, or congenital are reported in the literature; in nasal reconstructions, contradictions and distinct techniques exist that can be argued. Using the literature, we observe these distinct techniques that can be surgery or in surgery to reestablish and to integrate the patient with satisfaction in the society.

Key Words: Rhinoplasty, nasal reconstruction, island flap, nasal prosthesis

Surgical excision of tumors from the face may create a defect that is difficult to restore. Skin grafts can only cover superficial defects and have a natural tendency to contract and may not take properly. In addition, because of color mismatch, it is not cosmetically identical to the face.

A considerable number of people each year acquire facial defects as a result of malignant disease, trauma, and congenital deformity. Malignancies of the nasal vestibule are rare and account for only 9% of all cancers of the nasal cavity. Nasal defects commonly seen by plastic surgeons result from trauma, burn injury, or tumor resection. Although nasal reconstruction is one of the oldest plastic surgery endeavors, techniques continue to evolve and be modified.

Full-thickness nasal defects involving the alar rim require the restoration of the skin, the cartilage, and the nasal mucosa. The nasal skeleton is the supporting structure of the nose, and its integrity must be either maintained or restored for successful reconstruction. Numerous flaps were proposed in full-thickness defects. This reliable flap represents an alternative technique of composed grafts, different nasobialal flaps, forehead flap, and so on.

Reconstruction of extensive nasal defects including the nasal bone, the septum, and the aesthetically defined units of the nose always presents with certain difficulties. Forehead and free flaps can be used for reconstruction of extensive nasal defects.

The septal nasal perforation is an important problem for the laryngologists and plastic surgeons. The reasons of septal nasal perforations are injuries, neoplasms, self-mutilation, chronic rhinitis, allergy, Wegener granuloma, sarcoidosis, tuberculosis, toxic metals (arsenic and chrome), some drugs (steroids), narcotizing agents (cocaine), and complications after endoscopic and septal nasal operations.

The main reasons that urge the patients to seek help are plastic cosmetic and functional impairments such as nasal obstruction. Surgical removal of the hyperplastic tumor mass is the treatment of choice for rhinophyma.

Prosthetic rehabilitation is a surgical alternative in functional-aesthetic facial reconstruction when conventional reconstructive surgery cannot be applied because of either the psychophysical conditions of the patient or an excessive substance loss. Prosthetic supply has been developed into a functional and aesthetic alternative to plastic and reconstructive surgeries.

The purpose of this study was realized in a review of the indexed English-language literature with the intention of coming up with solutions for reconstructive surgery and no surgery of nose defects.

DISCUSSION

Solutions for Reconstructive Surgery and No Surgery of the Nose Defects

Nasal defects are the most challenging of these facial defects. Nasal reconstruction requires reconstruction of 3 tissue layers: the inner mucosal lining, the supporting structures (the cartilage or the bone), and the outer cutaneous lining. The new inner lining should consist of a well-vascularized thin tissue to prevent rejection of the cartilage or the bone in the supporting layer. For example, septal mucosa or skin can be used in various ways to reconstruct the inner lining. The new supporting structures, consisting of transplanted cartilage or bone, should be strong enough to prevent contraction of the soft tissues’ inner and outer linings during wound healing. The outer lining is reconstructed per subunit of the nose, for example, the nostril, the ala nose, or the ridge of the nose. Usually, a paramedian forehead flap is used for the reconstruction of the outer lining.

Surgical treatment, especially in cases of large septal perforation, is often difficult because of atrophy of the nasal mucosa and lack of suitable material for reconstruction. In surgical treatment, many methods and reconstructive materials have been used. The following autogenous tissues were used in the reconstruction of septal perforation: alloderm, temporal fascia, septal and auricle cartilage, cranial perioSTEum, perichondrium, ethmoidal, and hip bone. The defect of such materials is progressive resorption. For many years, the suitable synthetic material for septal nasal reconstruction has been searched for. Among the biomaterials, the following have been used without success: Dacron, porous polyethylene, dolomite, and bioglass. The rejection of the synthetic material was the reason of the failure. In the implantation of the resorbable copolymer of glycolide and e-lactide, very good macroscopic and histological results were achieved in vitro.

To improve the function of the external airways, the patient underwent surgery to reconstruct the nasal septum. Although cartilage grafts are the state of the art to reconstruct the nasal septum, a bone graft from the iliac crest is used because the autoimmune polychondritis precludes cartilage grafting because of expected cartilage destruction. The use of bone grafts is a promising method to restore and improve ventilation disorders caused by a saddle nose deformity in relapsing polychondritis. The septum is reconstructed by insertion of a bone graft taken from the iliac crest. This method has been used and described in orofacial surgery before. However, it has been proven to result to bone resorption. Here, cartilage grafting is not advisable, and bone grafting is the method of choice.

Although autologous tissue is known as the best material, porous polyethylene (Medpor) can be used safely regarding its low complication rate because of its porotic architecture and low morbidity in donors. Medpor implant provides functional improvement by its strength body. Its smooth surface helps to correct the asymmetry.
It is assumed to be the end stage of the perichondrial cutaneous graft technique.

The most common malignant tumors in this report the use of prelaminated radial forearm flap with porous polyethylene implants for total nasal reconstruction.

Because of the aesthetic consequences that result from the facial skeleton bony structure resections and the contiguous vital organs proximity, as in the orbit and the brain. The most common malignant tumors in this area are chondrosarcoma and osteogenic sarcoma. Because of the bone limits of the paranasal sinus and the proximity of the vital structures, many times, the surgical reconstruction is not indicated, with a nasal rehabilitation with prosthesis being necessary (Figs. 1–5).

Facial reconstruction with free microvascular flaps only has rarely produced an aesthetic result. Menick stated that distant skin always appears as a mismatched patch within a residual normal facial skin. In addition, earlier techniques using a single large nasal lining flap or bilateral nasal lining vaults incurred a high incidence of airway obstruction. The authors used microvascular free flaps and have proved them to be highly reliable and efficacious for restoration of missing elements of the nasal lining and adjacent facial soft tissue defects in total and subtotal nasal reconstructions. Combined with a forehead flap, this aesthetic approach allows for reconstruction of the center of the face layer by layer and facial unit by facial unit. Specific attention is paid to the artistic creation of normal nasal dimensions, proportion, and form using carved and assembled cartilage grafts and by secondary subcutaneous contouring. In addition, this technique produces a patent airway. Rhinophyma, the end stage in the development of acne rosacea, is characterized by sebaceous hyperplasia, fibrosis, follicular plugging, and telangiectasia. It is assumed to be the end stage of chronic acne rosacea.

Although it is commonly considered a cosmetic problem, it can result in gross distortion of soft tissue and airway obstruction. Basal cell carcinoma is a rare finding in patients with rhinophyma. The patient experienced chronic drainage and recurrent infections that failed conservative treatment with oral and topical antibiotics. The patient decided to proceed with surgical intervention and underwent tangential excision and dermabrasion in the operating room.

Absence of half of the nose is an extremely rare congenital malformation, which has a devastating impact on the patient and the family. Pathogenesis is not clear, and the reported cases have

![Nasal defect.](Image)

The functional and the aesthetic outcomes were satisfactory in most patients in whom a helical rim flap was used. The free vascularized preauricular and the helical rim flaps are a reliable method of reconstructing nasal defects and have wide clinical applications.

For Kalbermatten et al., the perichondrial cutaneous graft technique provides a reliable composite graft routinely harvested from the anterior conchal bowl. This established perichondrial cutaneous graft was simplified by using the less conspicuous posterior auricular donor site, which can be closed without the need of cartilage ressec- tion for reconstruction with a postauricular interpolated skin island.

The surgical conduct for these cases is very complex because of the aesthetic consequences that result from the facial skeleton bony structure resections and the contiguous vital organs proximity, as in the orbit and the brain. The most common malignant tumors in this area are chondrosarcoma and osteogenic sarcoma. Because of the bone limits of the paranasal sinus and the proximity of the vital structures, many times, the surgical reconstruction is not indicated, with a nasal rehabilitation with prosthesis being necessary (Figs. 1–5).

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sporadically occurred. Many aspects must be considered when reconstructing a congenital half-nose, such as the timing of surgery, the type of tissue to be used, and the need to reconstruct the nasal airway. Nasal reconstruction was performed at the age of 5 to 7 years to minimize psychologic trauma. Forehead skin demonstrated to be an excellent donor site to resurface the nose. For the inner lining, a contralateral cutaneous nasal flap was our preference. Concerning the nasal framework reconstruction, alar contour was restored using a cartilage graft from the lower portion of the ear tragus and the concha.

Nasolabial flaps are very useful and versatile local flaps, with robust vascularity that can be readily elevated without a delay. The flap can be superiorly based to reconstruct defects on the cheek, the side wall or the dorsum of the nose, the alae, the collumula, and the lower eye lid. The orbicularis oris myomucosal island flap is based solely on part of the orbicularis oris muscle as a new flap for nasal lining. The flap is designed horizontally on the oral mucosa of the upper lip along the gingivolabial sulcus. The upper margin of the flap is designed just below the buccal sulcus. The pedicle location is selected based on the location of the nasal lining defects. The medial muscle pedicle is chosen when the defect is located behind the columella and the soft triangle, the lateral muscle pedicle is chosen when the defect is located around the lateral two thirds of the ala, and the bilateral or unilateral muscle pedicle is chosen when the defect is located around the nasal floor.

For immediate nasal reconstruction with superficial temporal fascia, open rhinoplasty was used for absolute removal of the absorbed foreign body from beneath the skin of the nasal dorsum by the use of a rasp or curette. Any remnant foreign body then was irrigated by an isotonic sodium chloride solution. It was incised and dissected in the temporal area at 3 cm to check the superficial temporal fascia. Fascia was harvested (7 × 3 cm) from the temporal area. The fascia was used to wrap the thin silicon implant, after which the fascia and implant were sutured using an absorbable suture material. The silicon implant wrapping with superficial temporal fascia was inserted into the depressed nasal dorsum to correct the nasal deformity.

Efficient prosthetic supply in the exposed facial area requires a well scheduled and close cooperation between the surgeon and the anaplastologist. Preoperative diagnostic evaluation and operative filling of the magnetic fixation in cooperation with the anaplastologist provide satisfying functional and cosmetic prosthetic outcomes for the patient. Advantages of this procedure combine early cosmetic rehabilitation with the option to directly investigate the tumor site after resection. Novel magnet systems provide a high degree of flexibility combined with confident fixation. Prosthetic rehabilitation should be offered and discussed with patients ahead of cancer surgery in the head and neck area. This procedure should be considered in particular in patients who reject or are not feasible for multistep reconstructive surgery.

The association of dental implants and prostheses promotes aesthetic and function in the patient with nasal defects. No patient experienced severe inflammation requiring administration of systemic antibiotics or surgical revision. Implant success was 100% in both the irradiated and the nonirradiated patients. The dental implants result in a high rate of success in retaining midface prostheses and offer good stability and aesthetic.

![FIGURE 2. Free flap transfer to the nose.](image_url)

![FIGURE 3. Patient’s face with nasal carcinoma.](image_url)

![FIGURE 4. Patient’s face with defect after total rhinectomy.](image_url)

![FIGURE 5. Nasal prosthesis.](image_url)
The titanium implants placed failed to support a nasal epithelium in a patient with hepatitis C virus, with an important parodontal disease, who experienced a postinfective necrosis of the nose after a liver transplantation; it was necessary to place an adhesive prosthesis. An implant failure was also observed in diabetic patients with extensive midfacial defects due to a mycotic infection, but it did not compromise the retention of the prosthesis. According to our experience, the indication to epithesis is when the conventional reconstructive interventions are inapplicable.12

The functional and aesthetic satisfaction of the patient after a nasal reconstruction surgery or with prosthesis is a great challenge of the studies in literature.

REFERENCES
25. Seth R. Thaller, MD, DMD*

Excision and Grafting of Palmoplantar Keratoderma

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Abstract: Palmoplantar keratodermas may present to the clinician with an extremely broad series of clinical findings. Management has also taken on a wide variety of medical and surgical modalities. The literature seems to provide evidence that optimum management consists of surgical excision with skin grafting. It is believed that this will eliminate all of the underlying tissue and associated skin appendages, which are believed to be the source of this abnormal skin entity. We present a case of a patient in which tangential excision with delayed split-thickness skin grafting was performed after initial application of an acellular dermal matrix (Integra). Unfortunately,
there was nearly immediate recurrence of this disease, and we, therefore, suggest a more aggressive approach to the initial excision.

**Key Words:** Palmoplantar keratodermas (PPK), split-thickness grafting

**CLINICAL REPORT**

A 5-year-old African-American girl presented to the plastic surgery clinic with a history of progressive skin thickening on the volar aspects of the hands and the plantar aspects of the feet since birth. At age 1 year, these areas became increasingly symptomatic with increasing pain on ambulation, writing, and with the use of utensils. There is no associated family history of similar skin changes.

Physical examination revealed areas of focal hyperkeratosis, consisting of yellow-brown plaques involving the pressure points of the plantar areas and smaller hyperkeratotic plaques on the volar aspect of the fingertips (Fig. 1). Flexion contractures were noted on the interphalangeal joint of the thumbs and the distal interphalangeal joints of the index fingers (Fig. 2). Narrowing of the fingertips was noted on the thumb and index fingers bilaterally. Prominent lunulae of the fingernails were noted. Medial displacement of the right great toenail was also present and coincided with the thickest area of plantar keratoderma. The most prominent area of palmar keratoderma was present on the right thumb, which also exhibited the most severe distal tapering.

Complete blood cell count and metabolic and lipid panel values were all within normal limits. Medical treatment was initiated with 10-mg etretinate daily with minimal improvement. The patient was referred to plastic surgery service for evaluation and treatment of the plantar keratoderma.

To facilitate postoperative ambulation, the initial procedure was confined to the right foot. Tangential excision of 2 hyperkeratotic plaques was performed to the depth of the deep dermis (Fig. 3). The plaque on the metatarsal/instep area measured 8 × 10 cm, and the plaque on the calcaneal area measured 3 × 4 cm. A dermal regeneration template consisting of cross-linked bovine cartilage and glycosaminoglycan matrix (Integra Life Sciences, Plainsboro, NJ) was applied to the wound and subsequently grafted with a thick split-thickness skin graft (Fig. 4) and vacuum-assisted closure.

Histopathologic examination revealed acanthotic and markedly hyperkeratotic squamous epithelium consistent with keratoderma.

**DISCUSSION**

Palmoplantar keratodermas (PPKs) exhibit a broad range of clinical characteristics possessing marked overlap between a variety of clinical entities. Itin and Fistarol1 presented a scheme for classifying these disorders based on genetic transmission and described a diffuse versus circumscribed pattern. However, they reported that the clinical spectrum of PPK may be interindividually variable. More recently, these disorders have been separated by elucidating their molecular and genetic defects.

We report a 5-year-old African-American girl with severe focal PPK present since birth with no associated family history of the disease. Presence of flexion contractures, prominent lunulae, tapering, and constrictions of the digits in association with PPK are suggestive of the diagnosis of Huriez syndrome, an autosomal dominant syndrome characterized by diffuse sclerodactyly, PPK, and nail changes.2 Reports of Huriez syndrome typically describe a mild diffuse and smooth PPK with atrophy of the fingertips and toes.3,4 Additional features such as flexion contracture, tapering of the digits, and hypoplastic nails with ridging and prominent lunulae have also been described.5,6 Histologic findings in Huriez are nonspecific. They are characterized by hyperkeratosis and acanthosis.4,7 Immunohistologic and ultrastructural studies performed on 2 patients revealed almost complete absence of Langerhans cells.5,7 Autosomal dominant genetic transmission has been reported in all cases of Huriez syndrome.3 Lack of family history and the
possible that this may be the first African American described with this syndrome indicate that our case may be considered unique. In addition, severity of the focal, thick hyperkeratotic plaques in our patient markedly contrasts with the diffuse/mild keratoderma described in previously reported cases.

Treatment of PPKs consists of topical keratolytics, systemic retinoids, and, in severe cases, tangential or full excision with skin grafting. Owing to the overwhelming thickness of the plantar hyperkeratosis that resulted in pain upon ambulation, our patient underwent excision with split-thickness skin grafting after failing treatment with etretinate.

There are 9 previously reported cases of plantar keratoderma treated by surgical excision with skin grafting. Although Lucker and Steijlen reported a case of full-thickness excision, no follow-up information was given. Wynn-Williams and Dencer each report 1 patient with a successful outcome and describe their procedures as full-thickness excision and complete excision, respectively.

Three patients underwent initial excision to the dermal level with subsequent grafting and experienced recurrence of the lesions. Two of these 3 patients underwent a second procedure in which the excision was carried down to the plantar aponeurosis with no recurrence reported upon 6-month and 5-year follow-up. Another report of total excision of the sole to the level of the aponeurosis was also reported to result in a successful outcome.

Bedard et al reported 2 patients who experienced recurrences in the grafted areas or at the periphery of the grafts. Excision was carried to the level of the deep subcutaneous fat. Results were nevertheless viewed as successful with functional improvement despite the need to treat focal recurrences with shave excision and etretinate.

In our patient, we chose to excise the lesions down to the dermal bed. Immediate recurrence and lack of graft viability were possibly due to insufficient depth of excision. Friction may have played a role in failure of graft take.

CONCLUSIONS

We present a case of PPK treated with excision and grafting. Previous case reports suggest that to prevent recurrence, excision of plantar keratoderma must be carried to the level of the aponeurosis, removing all skin appendages. However, caution is advised when treating children, as one must take into account the risk of growth restriction if a graft is placed directly on the plantar aponeurosis. Lack of response to medical therapy in addition to these considerations would lead clinicians to continue to seek effective treatment modalities.

REFERENCES


Supraorbital Nerve Neuroma Caused by Blind Curettage of an Infected Epidermal Cyst

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Abstract: A 38-year-old woman presented with a tender mass with mild erythematous change above the left eyebrow area. She had received curettage in another clinic after a diagnosis of infected epidermal cyst 5 years previously. On examination, a round, irregular scar and a mass of 1-cm diameter associated with mild erythematous region were observed above the right eyebrow.

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Exploration disclosed a 1-cm neuroma with mild adhesion to a branch of the supraorbital nerve. The neuroma was completely removed. However, although partial transection of the supraorbital nerve was performed during surgery, the nerve was not repaired. The open wound was repaired securely. A histopathologic examination of the resected specimen resulted in a diagnosis of traumatic neuroma. The patient had no forehead numbness before or after surgery.

**Key Words:** Supraorbital nerve, neuroma, curettage

**DISCUSSION**

Traumatic neuroma may result from partial laceration, complete transection, or external compression by acute nerve injury or chronic nerve entrapment. Furthermore, axonal injury initiates a reparative response, whereby the distal portion of the damaged axon is gradually absorbed in a process called Wallerian degeneration and the proximal portion retains a potential for regeneration. If displacement has occurred between the nerve endings, sprouting axons may be unable to reach the distal endoneural tubes and will instead continue to elongate and proliferate at the site of the injury to form a neuroma. In this paper, the authors document a case of supraorbital nerve neuroma caused by simple and blind curettage of infected epidermal cyst above an eyebrow.

**CLINICAL REPORT**

A 38-year-old woman noticed a tender mass with mild erythematous change above the left eyebrow. She had undergone curettage in this region at another clinic for an infected epidermal cyst 5 years previously. She had visited our hospital because the lesion had recurred at the original site. On examination, a round 1-cm-diameter subcutaneous mass was found at the original irregular scar with an associated 1-cm-diameter mild erythematous mass above the left eyebrow (Fig. 1). She complained of mild tenderness and cosmesis, but she had no numbness of the forehead or tingling sensation. Our preoperative impression was that the mass was a recurrence of the previous epidermal cyst.

**FIGURE 1.** A 38-year-old woman noticed a 1-cm tender mass with mild erythematous change above the left eyebrow area (arrow).

The subcutaneous mass was identified under a loupe as a 1-cm-diameter neuroma during operation, which mildly adhered to a branch of the supraorbital nerve. The neuroma was completely removed, and although partial transection of the supraorbital nerve was performed during surgery, no repair was conducted. The open wound was repaired securely. The subsequent examination of the resected mass revealed it to be round, irregular, and 1 cm in diameter (Fig. 2). Disordered nerve fiber bundles intermixed with connective tissue. The small bundles of axons were surrounded by organized layers containing Schwann cells, fibroblasts, and perineurial cells (Fig. 3). Accordingly, a diagnosis of traumatic neuroma was made. The patient had no numbness of the forehead before or after surgery. During 1-year follow-up, the operative scar was acceptable and no complications were found, such as hypoesthesia and recurrence.

**FIGURE 2.** The mass was irregular, firm, and 1 cm in diameter.

The supraorbital nerve is a branch of the frontal nerve, which originates from the ophthalmic branch of the trigeminal nerve. This nerve runs through the supraorbital foramen or notch, ascends the forehead with the supraorbital artery, and then divides into a smaller medial and lateral branch, which supply the skin of the scalp almost as far back as the lambdoid suture. The supraorbital nerve typically exits the frontal bone via a palpable notch or foramen along the medial third of the supraorbital rim. However, in rare instances, this exit may be located anywhere along or above or even lateral to the supraorbital rim. Immediately after the supraorbital nerve exits its foramen, the nerve trunk forms superficial (medial) and deep (lateral) divisions. The superficial (medial) division passes over the frontalis muscle and provides sensory supply to the forehead skin and the anterior margin of the scalp. On the other hand, the deep (lateral) division runs cephalad to the upper part of the parietal bone, and provides sensory innervation to the frontoparietal scalp. When blind curettage is performed around the supraorbital foramen, injury of this superficial division may cause a neuroma to...
Recurrent and Refractory Giant Cell Tumor of the Jaw

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Abstract: Recurrent giant cell tumors refractory to various treatment modalities are challenging dilemmas for the most experienced practitioner. We report a case of a multiply recurrent aggressive giant cell tumor diagnosed at 10 months of age with extensive involvement of the maxillae and mandibles unresponsive to multiple therapeutic modalities. The various treatments attempted in this patient including conventional therapies as well as the original use of bevacizumab (Avastin) are described.
Clinical photograph of the patient after 2 resections and 3 months of interferon therapy (June 2004).

Although a number of tumor types may demonstrate large numbers of giant cells, 3 distinct entities are found in the head and neck—brown tumor of hyperparathyroidism, giant cell reparative granuloma, and true giant cell tumor. In 1986, giant cell tumors were classified into 2 subgroups—nonaggressive and aggressive based on a clinicopathology study of tumors arising in the maxillofacial skeleton. Nonaggressive tumors included brown tumor and giant cell granuloma and were defined by slow growth, minimal symptoms, and a low rate of recurrence. Aggressive tumors, the “true” giant cell tumors of the jaw, were defined as greater than 5 cm, rapidly growing, recurring, or resulting in tooth displacement, root resorption, or cortical perforation. Given the virtually identical histological appearance of giant cell lesions, this stratification allowed for treatment based on clinical presentation.

Our patient was classified as having an aggressive giant cell tumor of the jaw based on her initial presentation. As we will discuss below, she had persistence of her giant cell tumors despite a temporary response and disease stabilization with various treatment modalities that included surgery, embolization, antiangiogenic therapy, chemotherapy, hormonal therapy, and bisphosphonates.

CLINICAL REPORT

An 8-month-old female infant presented in June 2003 with a 5-month history of gingival hyperplasia and bleeding gums. Medical history was significant for a neonatal intensive care unit stay secondary to chronic lung disease and failure to thrive. She also had a history of osteoglophonic dwarfism, cranial vault remodeling, tracheostomy, and gastrostomy-tube placement.

A facial computed tomographic (CT) scan was obtained that demonstrated a soft tissue mass involving the maxilla and mandible. Intralesional biopsy confirmed giant cell tumor pathology. She underwent 2 resections by the age of 1 year, the first of which included administration of intralesional steroids and the extraction of 2 teeth. Because of the rapid tumor growth, she was given interferon α2b therapy in March 2004. After a period of slower growth, the tumor rapidly began to increase in size (Fig. 1). In addition, her interferon therapy was associated with intractable vomiting and marked electrolyte abnormalities. Interferon therapy was therefore discontinued after only 3 months. Two additional attempts at tumor debulking resulted in significant blood loss and infection. A subsequent CT scan demonstrated impending invasion of the cranial base. Chemotherapy with doxorubicin and cyclophosphamide was therefore initiated in an attempt to decrease the tumor size and to divert it away from vital cranial base structures. The patient received 6 courses of chemotherapy with the same agents between July 2004 and December 2004 and achieved a partial response. Therapy with pegylated interferon α2b was initiated on a weekly basis during the last 3 months, with the hope that it would contribute to her continued response and have fewer adverse effects than previously. Chemotherapy was discontinued because of the fear of cardiotoxicity associated with cumulative anthracycline dose. Shortly after this, the tumors progressed, prompting another embolization and an additional 2 resections. Despite these resections, the tumors recurred and continued to increase in size. Bisphosphonate therapy with intravenous pamidronate therapy was initiated, together with the ongoing interferon therapy. During this period, progressive tumor growth resulted in an additional embolization and 2 further debulking procedures. In March 2006, the interferon therapy was discontinued secondary to ineffectiveness and continued tumor growth, but pamidronate therapy was continued to improve the poor bone health associated with osteoglophonic dwarfism. The patient was then administered a low-dose vincristine intravenously and cyclophosphamide enterally as antiangiogenic therapy. This, too, was stopped 5 months later secondary to ineffectiveness and tumor-associated bleeding.

In August 2006, an interdisciplinary conference was held with the patient’s family regarding further treatment options. A trial of calcitonin administered subcutaneously was offered. Continued growth of these tumors prompted 3 more embolizations and debulking procedures and discontinuation of the calcitonin. Computed tomographic scans continued to demonstrate enlarging tumor. Because of the recalcitrant nature of the tumor, the patient was given a trial of bevacizumab (Avastin [Genetech, Inc, South San Francisco, CA]).

Key Words: Giant cell tumor, mandible, maxilla, Avastin, bevacizumab, antiangiogenic therapy, osteoglophonic dwarfism

FIGURE 1. Clinical photograph of the patient after 2 resections and 3 months of interferon therapy (June 2004).

FIGURE 2. Coronal three-dimensional CT reconstruction (December 2008).

FIGURE 2.
Thalidomide and one of its gene, 2 sagittal three-dimensional CT reconstruction (2009 Mutaz B. Habal, MD) therefore, a late-stage tumor is likely directly related to the comorbidity of osteoglophonic dwarfism. Osteoglophonic dwarfism is caused by activating mutations in FGFRI, a receptor for fibroblast growth factors (FGFs) that is presumably involved in the modulation of bone elongation. This similar resistance to therapy is not unexpected that our patient should have such an aggressive form of bone tumor.

What is very disturbing about this case is the patient’s level of unresponsiveness to antiangiogenic therapies. Resistance to antiangiogenic therapy in cancer is postulated to be the result of several factors. Some tumor cells demonstrate an intrinsic resistance to antiangiogenic-induced hypoxia, perhaps due to a mutant p53 gene, and are therefore less affected by hypoxia-inducing therapies. Another mechanism of resistance is the use of existing blood vessels (cooption) by tumors growing in vasculature-rich areas. More importantly, VEGF and its regulators seem to have a maximum effect in early angiogenesis and tumor development. Later angiogenesis is importantly, VEGF and its regulators seem to have a maximum effect in early angiogenesis and tumor development. Later angiogenesis is under the control of additional factors, including basic fibroblast growth factor and platelet-derived growth factor (PDGF). Therefore, a late-stage tumor might escape anti-VEGF therapy by upregulating these additional factors in response to antiangiogenic-induced hypoxia. The role of all these factors in giant cell tumors of bone is still unclear.

Recently, it has been found that monoclonal inhibition of the PDGF receptor results in decreased vascularity and tumor growth of end-stage tumors. Newer angiogenic inhibitors (ie, sorafenib, sunitinib) that target both VEGF and PDGF receptors are associated with loss of a previously stable vascular bed. Given their ability to target angiogenic receptors at various stages of vessel growth, sorafenib and sunitinib have recently been approved as monotherapy for renal cancer, and sunitinib has been approved for use in gastrointestinal stromal tumors. Unfortunately, these agents are available only in an oral preparation, precluding their use in our gastrostomy-dependent patient.

For our patient, other antiangiogenic therapies such as thalidomide might be beneficial. Thalidomide is a powerful teratogen, and in utero limb deformities are now considered to be the result of thalidomide’s ability to decrease secretion of both VEGF and basic FGF, thereby inhibiting angiogenesis. Thalidomide and one of its newer structural analogs, lenalidomide, have recently been approved for the treatment of multiple myeloma. Thalidomide and especially
lenalidomide have also proved efficacious in the treatment of myelodysplastic syndrome. Their use is currently under investigation in the treatment of other solid tumors, including Kaposi sarcoma and malignant melanoma. Therapeutic use of thalidomide and its derivatives is restricted and requires enrollments in safety programs designed to prevent possible teratogenic outcomes. Similar to sorafenib and sunitinib, thalidomide is available only as an oral preparation. Administration of thalidomide to our patient would be particularly difficult, as thalidomide would have to be crushed for gastrostomy-tube administration, thereby potentially exposing caregivers of childbearing age to its teratogenic effects.

Giant cell tumors of bone are also rich in RANKL (receptor activator of nuclear factor κB ligand), which plays a key role in bone destruction in several diseases that affect bone. Recently, denosumab, a fully monoclonal antibody against RANKL, was reported to have caused responses in 87% of adult patients with unresectable or refractory giant cell tumors. Unfortunately, this agent is not available to patients in the pediatric age group at this time even on a compassionate basis.

This patient represents the first reported case of a giant cell tumor of the jaw that has not responded to any available treatments. Continued treatment of this patient remains challenging and unprecedented. Further research needs to be conducted to elucidate the mechanisms of giant cell tumor formation in osteoglophonic dwarfism that may provide insight for potential therapeutic approaches. In addition, understanding current antiangiogenic therapies and their role in giant cell tumors will remain critical to the treatment of this condition.

REFERENCES

Use of Wide Bipedicled Pericranial Flap in Anterior Scalp Reconstruction

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Abstract: Pericranial flap is a composite flap involving the periosteum of the skull with its overlying loose areolar tissue termed subgaleal fascia. The multiple blood supply of the pericranial tissue enables this versatility, with a rich, anastomosing arterial supply from the supraorbital, supratrochlear, superficial temporal, posterior auricular, and occipital vessels. Thus, the shape, size, and location of the pericranial flap could be altered as long as a sufficient pedicle width could be fashioned to maintain a blood supply. In our study, we have performed wide bipedicled pericranial flap in scalp reconstruction in 2 cases. After tumor excision was completed, a pericranial flap was planned on the caudal side of the defect. A bipedicled-based pericranial flap was outlined with the use of a sharp dissection; this flap was elevated in a submusculoaponeurotic plane. The bipedicled pericranial flap, whose arterial supply was from the superficial and posterior auricular arteries, was transposed to the frontal defect.

We preferred a bipedicled flap, whose arterial supply is from the superficial temporal and posterior auricular arteries to augment vascular supply. If a large, long pericranial flap is required, making the flap pedicled ensures stable blood supply.

Key Words: Bipedicled pericranial flap, wide defects, stable vascularity

Pericranial flap is a composite flap involving the periosteum of the skull with its overlying loose areolar tissue termed subgaleal fascia. The anatomy of the pericranial flap has been reported by Potparic et al, who described subgaleal fascia as several thin connective tissue layers that are loosely adherent and glide over one another. The multiple blood supply of the pericranial tissue enables this versatility, with a rich, anastomosing arterial supply from the supraorbital, supratrochlear, superficial temporal, posterior auricular, and occipital vessels. Perforating vessels from the supraorbital, supratrochlear, occipital, posterior auricular, and superficial temporal arteries have been designed based on 2 different axial pattern blood supplies: anteriorly and laterally. The anteriorly based flap is supplied by deep perforating branches of the supraorbital and supratrochlear arteries, whereas a laterally based flap is supplied by the branches of the superficial temporal artery. Thus, the shape,
size, and location of the pericranial flap can be altered as long as a sufficient pedicle width can be fashioned to maintain a blood supply.\textsuperscript{2,3} Takagi et al.\textsuperscript{4} harvested bipedicled pericranial flap, whose arterial supply was based on superficial temporal and occipital arteries, for dura mater and cranial bone reconstruction. We did not find in the literature bipedicled pericranial flap designs whose arterial supply is based on superficial temporal and posterior auricular arteries. In our study, we have performed wide bipedicled pericranial flap in scalp reconstruction in 2 cases. We preferred bipedicled flap whose arterial supply is from the superficial temporal and posterior auricular arteries to augment vascular supply. If a large, long pericranial flap is required, making the flap pedicled ensures stable blood supply.

**MATERIALS AND METHODS**

After tumor excision was completed, a pericranial flap was planned on the caudal side of the defect. A bipedicle-based pericranial flap was outlined with the use of a sharp dissection; this flap was elevated in a submusculoaponeurotic plane. The tissues are dissected down to the pericranium and dissected to expose the desired width and length of the pericranial flap while the skin edges are retracted. The desired dimensions of the flap are outlined, and then the flap is cut from the caudal and cranial sides. The bipedicled pericranial flap, whose arterial supply was from the superficial and posterior auricular arteries, was transposed to the frontal defect.

It is important that the pericranial flap does not bear tension; because of this, it is then carefully sutured in place with interrupted sutures.

**Patient 1**

A 49-year-old male patient has squamous cell carcinoma since 2 years on the frontal region, and the lesion has invaded the frontal bone (Fig. 1A). The anterior table of the frontal sinus that has tumor invasion was outlined and removed using a 2-mm burr. At this point, the sinus mucosa was exenterated. After tumor excision was completed, a pericranial flap was planned on the caudal side of the defect; with the use of a sharp dissection, this flap was elevated in a submusculoaponeurotic plane. Flap size was 20 × 10 cm (Fig. 1B). The pericranial flap was used to cover the bony defect of the frontal sinus and exposed part of the frontal bone. So that the pericranial flap does not bear tension, it is carefully sutured in place with interrupted sutures, and then, split-thickness skin graft was adopted (Fig. 1C). Our follow-up period was 10 months, and in this period, we observed that graft intake was complete and frontal depression was minimal (Fig. 1D). We did not see wound dehiscence or partial or total flap necrosis.

**FIGURE 1.** A 49-year-old male patient has squamous cell carcinoma since 2 years on the frontal region (A). After tumor excision was completed, a pericranial flap was planned on the caudal side of the defect; with the use of a sharp dissection, this flap was elevated in a submusculoaponeurotic plane (B). So that the pericranial flap does not bear tension, it is carefully sutured in place with interrupted sutures, and then, split-thickness skin graft was adopted (C). Graft intake was complete, and frontal depression was minimal (D).
Patient 2

A 52-year-old male patient was admitted to our clinic because of recurrent basal cell carcinoma on the frontal region without bone invasion (Fig. 2A). Once tumor excision was initiated, periosteal involvement of the tumor was realized, but there was no bone invasion. After measurement of the defect size was completed, bipedicled pericranial flap was planned from the caudal side of the defect to cover the exposed bone. Flap size was \(5 \times 10\) cm (Fig. 2B). It is carefully sutured in place with interrupted sutures, and then, split-thickness skin graft was adopted (Fig. 2C). Our follow-up period was 1 year, and in this period, we observed that graft intake was complete and frontal depression was minimal (Fig. 2D). We did not see wound dehiscence or partial or total flap necrosis.

RESULTS

We have performed bipedicled pericranial flap on 2 patients who underwent tumor surgery. Our mean follow-up time was 11 months. Complete flap failure and full-thickness skin-graft necrosis did not occur; also, we did not see any wound infection or dehiscence.

Both patients stated complete satisfaction with the results. Neither patient reported tumor recurrence, and both were pleased with the aesthetic results and the frontal contour.

DISCUSSION

The term pericranium has been used to refer to the skull periosteum or to the composite of the periosteum and overlying subgaleal fascia. In our study, the periosteum and subgaleal fascia were included, making the flap more robust and easier to harvest than a pure subgaleal fascial flap. Pericranial flaps are used by all surgical specialties that might involve the head and neck. The range of applications for which pericranial flaps have been used is as varied as the specialties that make use of it. Wolfe reported the use of a pericranial flap to support rib grafts in cranial reconstruction. Common uses include the reconstruction of skull-base defects, frontal sinus obliteration, and the closure of abnormal communications between the orbit, sinuses, and nasal cavity.

We have used it mainly as a vascularized base for full-thickness skin grafting where bone has been exposed by excision of tumors extending to the periosteum. By including both periosteum and some subgaleal fascia in the flap, it also has some bulk, helping to fill out depressions that may otherwise make the repaired defect distinct and unsightly compared with the surrounding normal tissues, as would be the case if split-thickness skin grafts were used. This well-vascularized versatile flap is easy to harvest and avoids second donor site and its associated morbidity. Also, secondary graft contracture was minimal because of the rich vascular supply of the bipedicled pericranial flap.

In literature, anteriorly or laterally based pericranial flaps could be harvested. In our cases, supraorbital and supratrochlear artery circulations were broken down with tumor excision. However, the vascular supply of our bipedicled pericranial flap depends on the superficial temporal and posterior auricular arteries. Takagi et al harvested bipedicled pericranial flap for dura mater reconstruction, but the vascular supply of their flap was based on the branches of the superficial temporal and occipital arteries. Our bipedicled flap, whose arterial supply was from superficial temporal and posterior auricular arteries, differs from their flap by vascular supply.
Furthermore, even in cases such as our cases in which a large, long pericranial flap is required, making the flap bipedicled ensures stable blood supply. In our cases, there was absolutely no problem with blood supply, and the graft took completely. Our experience with the pericranial flap shows that it is a versatile, robust flap that is relatively easy to harvest. However, the pericranial flap does not bear the tension; because of this, it should then be carefully manipulated. This technique could provide good cosmetic results with minimal frontal depression and nice frontal contour.

REFERENCES

Nasopharyngeal Encephalocele: Report of Transcranial and Transpalatal Repair With a 25-Year Follow-Up

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Background: Encephaloceles are an extension or protrusion of any intracranial matter through a cranial bone defect. The defect is usually located along a cranial suture or at the junction of several bones. Most common locations of the defect are in occipital, orbital, or frontal areas. They reportedly occur in 1 to 3 of 10,000 live births. Encephaloceles extending into the nasopharynx may cause airway obstruction and feeding difficulty and present a potential pathway for central nervous system infection. Repair of the encephalocele should then be performed as soon as possible. Care of patients with nasopharyngeal encephaloceles requires a lifetime of reconstructive surgery. Care of these patients can be rewarding to both families and surgeons.

Encephaloceles are an extension or protrusion of any intracranial matter through a cranial bone defect. The defect is usually located along a cranial suture or at the junction of several bones. Most common locations of the defect are in occipital, orbital, or frontal areas. They reportedly occur in 1 to 3 of 10,000 live births. Encephaloceles extending into the nasopharynx may cause airway obstruction and feeding difficulty and present a potential pathway for central nervous system infection. Repair of the encephalocele should then be performed as soon as possible. Care of patients with nasopharyngeal encephaloceles requires a lifetime of reconstructive surgery. Care of these patients can be rewarding to both families and surgeons.

Key Words: Nasopharyngeal encephalocele, transcranial repair, transpalatal repair

Encephaloceles are an extension or protrusion of any intracranial matter through a cranial bone defect. The defect is usually located along a cranial suture or at the junction of several bones. Most common locations of the defect are in occipital, orbital, or frontal areas. They reportedly occur in 1 to 3 of 10,000 live births. The etiology remains unknown, although occasionally they have been observed to occur in families whose members have developmental anomalies of the central nervous system (CNS). Encephaloceles are divided into 2 groups according to the location of involvement: those involving the convexity of the skull and those involving the base of the skull. The latter represents only 10% to 15% of all encephaloceles. The sphenothmoidal encephalocele is of the basal group. It has been described as protruding into the nasal cavity through a defect in the sphenoid and ethmoid bones and extending farther into the nasopharynx via a palatal cleft. Such a nasopharyngeal encephalocele may cause airway obstruction and also represents a potential pathway for infection of the CNS. In addition, vital brain structures may be damaged if not reduced into the cranium. For those who survive long enough, expedient repair is critical. Of the few cases reported, repair has been described via a transcranial or combined transoral/transpalatal approach. Because the transcranial repair has been reported with a low success rate and mortality rate of 50%, some surgeons have advocated the latter approach. We report a case that was repaired successfully via a combined transcranial and transpalatal approach. Furthermore, because of his successful repair, the patient underwent multiple secondary reconstructions resulting in a 25-year follow-up.

CLINICAL REPORT

A 3960-g, term male infant, from a normal gestation, was delivered via cesarean delivery for breech position. Initial examination revealed a 2 × 3-cm gray intraoral nasopharyngeal mass with smooth
mucosal covering extruding through a midline palatal cleft. This mass was compressible and easily reducible into the cleft. Marked hypertelorism, a bifid nose, and a midline “O” cleft lip were also present. The rest of the physical examination was unremarkable.

Skull radiographs showed a midline defect of the ethmoid and sphenoid bones with absence of the cribriform plate, medial portion of anterior fossa floor, planum, and tuberculum. The sphenoid, sella turcica, and anterior and posterior clinoids were present. In addition, the computed tomography (CT) scan showed a soft tissue density mass in the nasopharynx, absence of the corpus callosum, and dilation of the posterior horns of the lateral ventricles (Fig. 1). The electroencephalogram and results of laboratory tests did not show abnormalities.

Hospital Course and Subsequent Treatments

Initially, the infant’s feeding was impaired because of the mass and palatal defect, and a feeding tube was placed. On the 14th day, the infant was found to have evidence of diabetes insipidus and a decreased level of serum adrenocorticotropic hormone. The patient was treated with vasopressin (Pitressin) and dextrose 5% in water. Twenty-four days after his birth, the patient developed respiratory distress secondary to airway obstruction by the nasopharyngeal mass. Emergent tracheostomy was performed.

At 4 weeks, extradural and intradural exploration of the encephalocele was performed via a bifrontal craniotomy. Intraoperatively, the encephalocele was found to be composed of a cystic cavity with copious, glistening, tough dysplastic tissue. Ninety percent of this tissue was removed. The inferior aspect of the encephalocele sac was lacking a dura mater and was thus essentially composed of arachnoid membrane and nasopharyngeal mucosa. The medial anterior cranial floor defect, extending from the ethmoidal to an area of anterior to posterior clinoid processes, was repaired with a bone graft from the bifrontal craniotomy. A dural graft was also required. Postoperatively, a cerebrospinal fluid leak was noted with subsequent development of Pseudomonas meningitis. Antibiotics were intravenously administered. At 6 weeks, a progressive nasopharyngeal swelling at the site of the repaired encephalocele was noted. The patient was found to have hydrocephalus by CT scan, and a ventriculoperitoneal shunt was placed. A follow-up CT scan showed diffuse air in the ventricular system with an air track between the nasopharynx and the apex of the frontal horn. At 10 weeks, it was noted that the repair had ruptured secondary to the development of hydrocephalus, and extradural and intradural exploration of the repaired anterior cranial floor was undertaken. A fistulous defect was found in the floor, which was repaired with cranial bone graft.
sandwiched between 2 sheets of lyophilized dura mater (Lyodura, Braun Melsungen AG, Marburg, Germany). In addition, transposition of a palatal mucosal flap (a right Millard island flap) was used to cover the inferior surface of the floor defect from the mouth. The hydrocephalus, pneumocephalus, and meningitis subsequently resolved, and the patient was discharged home at 13 weeks on DDAVP (1-deamino-5-d-arginine vasopressin) and hydrocortisone.

On subsequent follow-up, the patient was found to have deficiencies of growth hormone and thyroid function and therefore was administered growth hormone and levothyroxine (Synthroid). A frontal epidural abscess that communicated with the nasopharynx was drained and treated with intravenously administered antibiotics. The fistula spontaneously closed with the resolution of abscess.

At 10 months of age, the patient had stabilized from his prior surgeries and began to gain weight appropriately but remained with his unrepaired median cleft lip and palate (Fig. 2). At 15 months of age, the patient underwent median cleft lip repair by making corresponding markings from each oral commissure to the high points of the Cupid’s bow. An inverted “V” design was traced on the lip from the apex of the cleft that lies at the base of the columella to 2 corresponding points 3 mm medial to the high points of the Cupid’s bow. A 90-degree angle in the marking for excision was made approximately 3 mm above the mucocutaneous white roll on each side of the cleft, to give vertical length to the repaired lip and provide fullness of the vermillion in the midline tubercle (Fig. 3).

The patient was followed up at 4 years of age, which revealed an excellent result of the median cleft lip repair. The patient had persistent hypertelorism (Fig. 4) and nasomaxillary hypoplasia (Fig. 5). At 6 years of age, the hypertelorism was corrected by wedge resection of the frontal and nasal bones and medial mobilization of the orbits. A preoperative photograph depicts the severity of the hypertelorism that progressed at age 6 years (Fig. 6). Intraoperatively, the medial interorbital distance measured 38 mm as well as a large nasal defect at the site of the repaired encephalocele (Figs. 7 and 8). Rib grafts were obtained, and the remaining nasal bones were used as a composite cantilever graft to reconstruct the nose after 12 mm was resected from the frontal and nasal bones.

FIGURE 5. Lateral photograph at 4 years of age showing the severe nasomaxillary hypoplasia secondary to the wide cleft palate and nasopharyngeal encephalocele.

FIGURE 6. Intraoperative on table photograph showing the severe hypertelorism.

FIGURE 7. With the periorbita retracted, the nasal defect can be seen from the prior nasopharyngeal encephalocele.

FIGURE 8. Intraoperative photograph after coronal incision and release of the periorbita showing an interorbital distance of 38 mm.
The frontal cranial defect was also reconstructed with multiple split rib grafts (Figs. 11 and 12). The hard and soft palates were not repaired but were obdurated by a prosthesis. Photographs obtained at 4 months postoperatively revealed excellent correction of the orbital hypertelorism as well as an augmented nasal radix and dorsum (Fig. 13). Hospitalization occurred at 12.5 years for hydrocephalus secondary to malfunction of the ventriculoperitoneal shunt. The shunt was revised, and the patient was discharged home without further complication.

**Growth and Development**

At 4 months of age, the patient was noted to have a problem with his vision and hearing and was developmentally delayed. His weight and height were both below the fifth percentile. Laboratory data indicated hypopituitarism. At the age of 3 years, his weight and height continued to be below the fifth percentile. Examination of the eyes showed slight amblyopia in the left eye with a constant nystagmus and a disconjugate gaze. Audiometry showed a response at 80 dB. Sound field responses were similar to those of a younger child with low-normal responses through speech range. At 8 years of age, his weight and height were only at 50% of children 4.5 years of age. Despite the continuation of growth stunting, his mental development was comparable to that of children at his age. By 12 years of age, he attended seventh grade and was reported to be a good student. Follow-up at 13 years old revealed maintenance of the nasal dorsum correction, with the beginning of recurrence of the orbital hypertelorism associated with a growth spurt (Fig. 14); however, he remained relatively underdeveloped secondary to the hypopituitarism. At 25 years of age, the patient returned for follow-up visit (Figs. 15 and 16). Photographs were obtained and revealed maintenance of the nasal dorsum and correction of the hypertelorism. There were a moderate orbital dystopia and persistent naso-maxillary hypoplasia likely secondary to the prior encephalocele and use of the Millard rotational flap to close the intracranial defect. The patient and his mother were extremely satisfied with the reconstruction.

**DISCUSSION**

As illustrated in our case, these rare patients with a sphenothmoidal encephalocele are associated with the following facial and cerebral deformities: hypertelorism, bifid nose, cleft lip, cleft palate, and partial or complete agenesis of the corpus callosum. Depending on whether the pituitary was involved, the patient may present with evidence of hypopituitarism. The patient may also have visual...
problems due to optic nerve atrophy.\textsuperscript{3-6} Therefore, in addition to physical examination, patients should have a complete radiological study of the head (skull film and head CT), an evaluation of endocrine function, and an ophthalmologic evaluation.

Patients frequently present with feeding difficulties and evidence of airway obstruction due to the presence of the encephalocele in the nasopharynx. In addition, its presence also creates a potential pathway for CNS infection. For these reasons, reduction of the encephalocele and repair of the cranial floor should be undertaken as soon as possible.

Mortality due to bacterial meningitis is relatively high, especially for neonates (between 25% and 50%).\textsuperscript{7} Because of this fact, transcranial repair of a nasopharyngeal encephalocele has been the recommended approach. However, transoral and transpalatal repair under an antibiotic umbrella has been advocated because it allows for easy manipulation and reduction of the encephalocele without disrupting vital structures within the encephalocele.\textsuperscript{5} As seen in our case, the bases of the encephalocele were found to be without dura. No dura can be detached from the nasal mucosa for the reduction of the encephalocele. Intradural and subdural structures would have been contaminated with naso-oral flora, and the risk of infection would have been increased if a transpalatal approach was used. In addition, to repair the cranial floor and dural defects in these patients, either an autologous or alloplastic graft would be required. By introducing the graft via a naso-oral route, the risk for contamination would increase the risk of postoperative infection.

Transpalatal repair has been reported to be associated with less incidence of hypopituitarism because it avoids resection of cerebral structures within the encephalocele.\textsuperscript{5} However, often these patients with nasopharyngeal encephalocele already have panhypopituitarism and risking infection for a transpalatal approach would not be prudent in these patients.

Because anterior cerebral arteries have been found to deflect inferiorly into the encephalocele,\textsuperscript{4} prior study of intracephalocele content is wise to prevent vascular injury and subsequent brain ischemia. These patients with nasopharyngeal encephaloceles have immediate life-threatening medical and surgical emergencies associated with airway compromise and endocrine dysfunction. However,
Desmoplastic Ameloblastoma

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Abstract: Ameloblastoma is a rare benign odontogenic epithelial tumor characterized by abnormal cell growth, which easily infiltrates and destroys surrounding bony tissues. Clinically, it is mostly seen in the ascending ramus area, in men in their fourth and fifth decades more frequently than in women. One of the 3 clinical variants of ameloblastoma is desmoplastic type, with involvement of the maxillary arch more often than the mandible. Its histopathologic variant characterized by extensive squamous metaplasia, islands of tumor cells, and sometimes keratin formation is known as desmoplastic acanthomatous ameloblastoma.

The aim of this report was to present an unusual case of synphysis located desmoplastic acanthomatous ameloblastoma in a 56-year-old female patient, who was experiencing laryngeal carcinoma 2 years ago.

Key Words: Ameloblastoma, desmoplastic acanthomatous ameloblastoma, synphysis mandible, radiolucency

Ameloblastoma represents 11% of all odontogenic tumors and originates from the epithelium, which is involved in the formation of teeth.1 Potential epithelial sources include enamel organ, odontogenic rests, reduced enamel epithelium, and the epithelial lining of odontogenic, especially dentigerous cysts. The trigger or stimulus for neoplastic transformation of the previously mentioned epithelial sources is totally unknown.2 If the unicystic type is formed from the epithelial lining of a dentigerous cyst, it is called a mural ameloblastoma.1 Ameloblastomas with a slight predilection occur in men and develop more often in black Africans.1 Localizations of ameloblastomas are mostly seen (80%) in the molar-ramus region of the mandible; however, they may develop in the symphyseal area.1,2 In the maxilla, the molar area is more commonly affected than the anterior or premolar area.3 Although the ameloblastoma is mostly an intraosseous tumor, there are cases of the so-called extraosseous peripheral ameloblastomas found in the gingiva or buccal or palatal mucosa.3-5 These peripheral forms of ameloblastomas comprise approximately 1% of all ameloblastomas.3 The ameloblastoma occurs predominantly in the fourth and fifth decades (35–45 y).6 Although it may be found in young (age, 3 y) people and in individuals older than 80 years,1 Recurrence rates are higher in older patients and in those with multilocular lesions. Local recurrence, whether detected radiographically or histologically, may have a more aggressive character than the original tumor.1 There is a pronounced tendency for ameloblastomas to cause extensive root resorption and tooth displacement. The stroma of the ameloblastoma usually consists of relatively acellular loose connective tissue. Sometimes, it is more collagenous and may be frankly desmoplastic with small nests and strands of odontogenic epithelium. This has been described as desmoplastic ameloblastoma.7 Desmoplastic ameloblastoma was first described by Eversole et al in 1984.7 According to the World Health Organization’s histopathological typing of odontogenic tumors, desmoplastic ameloblastoma is a benign and local invasive jaw tumor, which represents approximately 4% to 13% of all ameloblastomas, with involvement of the maxillary arch more often than the mandible.6-12 The term acanthomatous is used with cases where there is extensive squamous metaplasia, sometimes with keratin formation, within the islands of tumor cells. The general pattern of the tumor is that of the follicular type. This variety must be distinguished from the squamous odontogenic tumor, in which the peripheral cells are flat rather than columnar.5

CLINICAL REPORT

In December 2008, a 56-year-old female patient referred to the Department of Oral and Maxillofacial Surgery, Istanbul University,
with a complaint of swelling in the region of anterior mandible, which was present for 2 years. Clinical intraoral examinations disclosed a very bad oral hygiene with periodontal disease, remnants of the teeth roots, and caries lesions of the most teeth. In the anterior part of the mandible from the vestibule side, a swelling was noticed, which was of soft consistency. The surface was smooth, and the color was similar with the adjacent mucosa. With appliance of pressure, white exudate was flowed from the fistula. There were no symptoms of pain or paresthesia. Teeth 41, 42, 43, 44, 45, 31, 32, 33, 34, and 36 associated with the lesion were nonvital. Extraoral examinations revealed a presence of asymmetry at the left side of the mandible, and the presence of lymphadenopathy at the left submandibular region was noted too. A laryngostoma was present, and the patient could not speak.

The radiographic examinations have revealed a large well-circumscribed radiolucent lesion that was 8 cm in diameter, located between the first right premolar and second left molar teeth, which was radiologically suspicious to be a radicular cyst (Fig. 1). The lower edge of the lesion was the inferior border of the mandible, whereas the upper edge in the left premolar region was in the level of oral mucosa and cervix of the lower left canine and the first premolar teeth.

From the patient's health history, it was learned that she had experienced laryngeal carcinoma. In the period between November 2003 to May 2005, the patient had undergone 3 different surgical procedures, which resulted to complete laryngectomy and anterior lymph node dissection. Having consulted the patient's specialist of oncology, it was found out that the patient had a squamous cell carcinoma of the larynx, which was treated just operatively because radiotherapy and chemotherapy were refused by the patient herself. Four months after the first operation (March 2004), the second one was performed, which resulted to complete laryngectomy and lymph node dissection as the consequence of tumor metastasis. In May 2005, a permanent laryngostoma was embedded. The patient was under follow-up for 4 years without any complaints and complications. Taking to consideration the general health condition of the patient, there were no contraindications to surgical intervention under local anesthesia because the surgery under general anesthesia was refused by the patient.

Before surgery, the patient underwent an endodontic treatment of teeth 41, 42, 43, 44, 45, 31, 32, 33, 34, and 36, which were involved in the cystic lesion. Under local anesthesia, a full mucoperiosteal, sulcular incision was made from the lower right first molar to the left
second molar. The mucoperiosteal flap was reflected into the fornix. At the beginning of operation, apicotomy with cyst enucleation was planned. However, during the curettage of the pathologic mass, the apicotomy was abandoned because there was no bone around the roots of the teeth. Therefore, the 31, 32, 33, 34, 35, 36, 41, 42, 43, and 47 teeth were extracted. The pathologic specimen was submitted to the Department of Oncologic Cytology and Tumor Pathology, Istanbul University, as the infected radicular cyst (Fig. 2). The flap was placed in its original position and sutured. After 8 days, the wound healed, and the sutures were removed. The patient reported a sense of numbness in the region of the lower lip and chin from the left side, which started a day after the surgery. The patient is followed up for 6 months, and there was no sign of infection or recidivation observed until now (Fig. 3).

According to histopathologic examinations, the lesion was diagnosed as acanthomatous desmoplastic ameloblastoma (Figs. 4–6).

**DISCUSSION**

Ameloblastoma is the second most common odontogenic tumor, which originates from the epithelial components of the tooth-forming apparatus. The stimulus for the neoplastic transformation of these tissues is not known yet.\(^6\),\(^7\),\(^13\)

Eversole et al.\(^1\) first described desmoplastic ameloblastoma in 1984, and their studies were later reviewed by Waldron and el-Mofty\(^10\) in 1987. These reports indicated that this type of ameloblastoma has many features that are inconsistent with conventional ameloblastoma. There is a dense fibrous connective tissue around the tumor island, a comparatively unusual location from other ameloblastomas, and a radiographic appearance that is similar to a benign fibroosseus lesion, which could be due to the infiltrative and osteoblastic behaviors of this tumor.\(^6\),\(^13\) Clinically, patients with typical ameloblastoma do not have significant initial complaints; however, in the cases of desmoplastic ameloblastoma, there are possibilities of initial pain.\(^14\)

If the central portions of the tumor islands became squamous or elongated, the terms *acanthomatous* and *spindle* are used to modify ameloblastomas. In our case, it was identified histopathologically:

![FIGURE 5. Tumoral masses surrounded by ameloblastic cells are observed (thin black arrows). These cells are in a connective tissue with dense collagen fibers (thick black arrows). There is also squamous epithelial metaplasia was seen at the middle of the quasi stellar cells (white thick arrows; [H&E], original magnification ×200).](image)

![FIGURE 6. Tumoral masses surrounded by ameloblastic cells are observed (thin black arrows). These cells are in a connective tissue with dense collagen fibers (thick black arrows). There is also squamous epithelial metaplasia was seen at the middle of the quasi stellar cells (white thick arrows; H&E, original magnification ×400).](image)
desmoplastic ameloblastoma was also located in the anterior mandible. Our case also showed this predilection for the anterior jaw.

REFERENCES

12. Philipsen HP, Reichart PA, Takata T. Desmoplastic ameloblastoma (including “hybrid” lesion of ameloblastoma). Biological profile based on 100 cases from the literature and own files. Oral Oncol 2001;37:455–460

Comparison of the Soft Tissue Thickness of the Midface in Craniosynostosis

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Abstract: Craniosynostosis is classified according to the included sutures as either symmetric, such as scaphocephaly and brachycephaly, or asymmetric, such as plagiocephaly. Asymmetric craniosynostosis has been known to exert asymmetric effects not only on the cranium but also on the facial skeleton. Nonetheless, the presence of asymmetry in soft tissues is only speculative and is based on the experience of plastic surgeons. In our study, after measuring the surface coordinates of soft and bone tissues from numerous positions on three-dimensional computed tomography (CT), the distance between the coordinates was calculated and the thickness of the soft tissues was obtained by objective measurements. The subjects included a control group of 8 symmetric craniosynostosis patients, and the study group consisted of 7 asymmetric craniosynostosis patients selected by computed tomographic data. In each patient, the thickness of the midfacial area was measured by dividing it into frontal and lateral views. The results show that in symmetric craniosynostosis, the thicknesses of the right and the left soft tissues were statistically identical. However, in asymmetric craniosynostosis, the thickness of soft tissues in the hypoplastic side was statistically significantly thinner than the measurements taken at both the frontal and the lateral views (P = 0.048 and P = 0.034, respectively). This suggests that surgeons should pay attention to the asymmetry of soft tissues during follow-up and when correcting facial asymmetry in asymmetric craniosynostosis.

Key Words: Craniosynostosis, soft tissue thickness, three-dimensional computed tomography

Craniosynostosis has different appearances depending on the location or number of synostosed suture lines, and it affects not only the cranium but also the midfacial area.1–3 According to
the included sutures, morphologic changes are shown as symmetric or asymmetric craniofacial skeletons. Particularly, plagiocephaly, which is caused by early closure of the unilambdoid or the unicoronal suture, induces asymmetry of not only the cranium but also the facial area. Such asymmetry represents all of the changes in the bone and the soft tissues. Nonetheless, asymmetry of the soft tissues has not been reported as the causative factor of asymmetry of the facial area, but asymmetry of bone tissues has been reported as such. This is considered to be due to the methods to measure asymmetry of soft tissues that effectively do not exist. In our study, the thickness of facial soft tissues was obtained by three-dimensionally reconstructing the images from computed tomographic (CT) scans, by measuring the coordinates on the surface of the skin and the surface of the bone tissues of the facial area from the predetermined same points, and by calculating the distance between the 2 coordinates. In addition, by comparing the thicknesses on the right and the left sides, the role of the soft tissues in the asymmetry of the midface in plagiocephaly was examined.

MATERIALS AND METHODS

Materials
The craniofacial CT scans of 15 patients with a diagnosis of nonsyndromic craniosynostosis were reconstructed as three-dimensional CT images using the Analyze AVW 5.0 (AnalyzeDirect, Overland Park, KS). The age range was 2 to 58 months, and 10 patients were boys, and 5 were girls. Among 15 patients, 8 had symmetric craniosynostosis (4 with sagittal craniosynostosis, 3 with bicoronal craniosynostosis, and 1 with metopic craniosynostosis) and 7 had asymmetric craniosynostosis (6 with unicoronal craniosynostosis and 1 unilambdoid craniosynostosis). Measurements were obtained from the symmetric craniosynostosis patients as the control group for comparison with those from the asymmetric craniosynostosis patients.

Methods
Axial and noncontiguous CT scans were obtained from the mentum to the vertex at 2-mm intervals. Three-dimensional reconstruction


FIGURE 2. Lateral view of the reconstructed three-dimensional images. Right (A) and left bone tissue accentuated images (B). Right (C) and left soft tissue accentuated images (D).
was performed using the Analyze AVW 5.0 software, and the pixel size was 0.5 mm. The reconstructed three-dimensional CT scan was realigned while the horizontal plane passed through the nasion and both porions, and the vertical plane formed a perpendicular angle with the horizontal plane and passed through the nasion and the basion. From the realigned three-dimensional CT scan, 2 types of bone tissue and soft tissue images were prepared while the density threshold controls. Subsequently, in the frontal and both lateral views of the soft tissue images, the x coordinates dividing the space between the right and the left tangent line to 4 equal parts and the y coordinates dividing the space between the highest and lowest tangent line to 9 equal parts were obtained. Based on the cross points of the x and y axes, x and y coordinates could be determined from the (x, y, z) coordinates. From the soft tissue images on the corresponding (x, y) coordinates, z coordinates were obtained, and similarly, the z coordinates of the bone tissue images were obtained; thus, the three-dimensional coordinates of the surface soft and the bone tissues of the same cross points were obtained. By calculating the distance between the soft and the bone tissue coordinates and applying the formula \[ d = \sqrt{(x_1 - x_2)^2 + (y_1 - y_2)^2 + (z_1 - z_2)^2}, \] the thickness of the soft tissues at each cross point was measured through multiplication by the pixel size (0.5 mm). Among each cross point, on the frontal view, 6 coordinates of the midface, excluding the coordinates in the midline, were acquired and termed ARm, ALn (n = 1–3; Fig. 1). On both lateral views, 12 coordinates of the 6 right and 6 left cross points corresponding to the midface, excluding the occipital, were selected and termed LLm, LRm (m = 1–6; Fig. 2). As a result, on the frontal view, the hypoplastic and the nonhypoplastic sides were compared according to the 3 pairs of cross points ARn and ALn (n = 1–3), and the 6 pairs of the right and the left cross points LLm and LRm (m = 1–6) in the lateral view were compared with each other statistically. For statistical analysis, \( t \) test \( (P < 0.05) \) was performed.

**RESULTS**

On the frontal view of the symmetric craniosynostosis group, the right and the left sides were measured (mean [SD]) at 29 (14) and 28 (16) mm, respectively; and on the lateral view, at 17 (17) and 17 (14) mm, respectively. The thicknesses of the soft tissues on the left and the right sides from the frontal and the lateral views were not statistically different \( (P = 0.440 \) and \( P = 0.936, \) respectively). On the frontal view of the asymmetric craniosynostosis group, the thickness of the hypoplastic side is significantly thinner than that of the nonhypoplastic side in the asymmetric craniosynostosis group. *The value is statically significant \( (P < 0.05) \). CRS indicates craniosynostosis.

<table>
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<th>Table 1. Comparison of Soft Tissue Thicknesses</th>
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<td>Group</td>
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<tr>
<td>Symmetric CRS</td>
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<td>Right, mean (SD), mm</td>
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<td>Left, mean (SD), mm</td>
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<tr>
<td>Asymmetric CRS</td>
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<td>Hypoplastic, mean (SD), mm</td>
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<tr>
<td>Nonhypoplastic, mean (SD), mm</td>
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<td>( P )</td>
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The thickness of the hypoplastic side is significantly thinner than that of the nonhypoplastic side in the asymmetric craniosynostosis group.

**DISCUSSION**

In the nonsyndromic craniosynostosis patients, the primary deformity of the cranium may secondarily induce deformity of the facial bones, and asymmetric deformities such as unicoronal synostosis and unilambdoid synostosis are commonly associated with hypoplasia of the bone tissues in the ipsilateral area. Recently, due to the generalization of cosmetic surgery, the interest of patients, guardians, and surgeons regarding the necessity of correcting facial asymmetry in congenital deformities has been on the rise. Therefore, it is important to more accurately understand facial asymmetry that may be developed in asymmetric craniosynostosis. However, study into the causes of facial asymmetry in craniosynostotic plagiocephaly patients has mostly been focused on the asymmetry of bone tissues, and there have been few studies that have examined the asymmetry of soft tissues.8,9 Measuring soft tissue thickness using three-dimensional CT scans in our study is a new method. By realigning three-dimensional CT scans on the standard horizontal and vertical plane, the location of deviation that may occur during CT was standardized, and the frontal and the lateral views among patients could be made identical. In addition, the cross points were selected by dividing the vertex and the mentum into 9 equal parts and the right and left sides into 4 equal parts. Therefore, the measurement points were determined under an identical demographic principle, which allowed us to minimize differences that may occur among patients. The distance between the coordinates on the surface of the skin and the bone from each cross point was defined as the soft tissue thickness. This method could not...
be a tool to measure the absolute soft tissue thickness, but it could allow for the best consistency and reliability compared with other methods reported until now, and it is meaningful as a noninvasive method to measure in vivo data.10–12

The results show that asymmetry of soft tissues in asymmetric craniosynostosis is also statistically significant as deduced by surgeons, which demonstrates that the asymmetry of soft tissues contribute partially to facial asymmetry in patients with asymmetric craniosynostosis. This suggests that attention must be paid during follow-up evaluation and corrective surgery. Considering that the importance and effectiveness of soft tissue reconstruction in the correction of hemifacial microsomia and other facial asymmetry has been reported, it is believed that similar concerns are warranted for asymmetric craniosynostosis.13,14

However, there were only 3 pairs of cross points in the frontal view in our study, which are considered to be insufficient. This is because our study subjects were pediatric patients; thus, the ratio of the mid-facial area, including the maxilla, was markedly smaller than that in adults. Therefore, it is assumed that such errors could be reduced by measuring more cross points through the subdivision of the area or the selection of already known landmarks in future studies.

CONCLUSIONS

Facial asymmetry developed in asymmetric craniosynostosis is accompanied not only by asymmetry of bone tissues but also asymmetry of soft tissues, which should be considered for the evaluation and treatment of this disease.

REFERENCES