Management of Aggressive Midface and Orbital Fibrous Dysplasia

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Objective: To discuss cosmetic and functional implications in the evaluation and treatment planning of large, aggressive midfacial fibrous dysplasias.

Patients and Methods: Eight patients (aged 2-38 years) with large fibrous dysplasias of the maxilla, zygomatic, and ethmoid bones requiring varying degrees of intervention and reconstruction were retrospectively reviewed. Patients with smaller lesions of these regions not requiring resection and reconstruction, as well those requiring sinus surgery alone were excluded from this review.

Results: All fibrous dysplasias in this review were monostotic, 6 originating in the maxilla, 1 in the zygoma, and 1 in the ethmoid. Five lesions (4 maxillary, 1 zygomatic) caused cosmetic deformity without functional deficits and required resection and/or contouring only with minimal reconstruction. The remaining lesions were invasive such that function of the eye and/or dentition was affected. These lesions were treated by aggressive resection and various degrees of reconstruction to optimize function.

Conclusions: While fibrous dysplasia is classified as a benign process, local expansion can cause significant functional and aesthetic deformities. Each lesion should be thoroughly evaluated and, when vital structures are involved or threatened, total or subtotal resection should be considered. A variety of options should be available to the surgeon for definitive primary reconstruction.


Fibrous dysplasia is the replacement of normal bone with fibrous tissue causing painless expansile lesions that impair cosmetic and structural function of bone. They constitute 7% of all nonmalignant bone tumors and may be either monostotic or polyostotic. The monostotic form is more commonly found in the facial skeletal region. These fibrous lesions are generally nonneoplastic, but rare malignant transformation has been described. Most of these fibrous lesions are believed to be self-limiting after puberty when the patient's growth is complete, but aggressive lesions in pediatric patients that involve the orbit, orbital apex, and skull base present significant management challenges.

These lesions tend to be a firm painless swelling or deformity of the maxillofacial region. The history of growth is gradual. Those lesions of the midface and periorbital region may be initially seen with nasal obstruction, sinus symptoms, headache, dental problems, and possible visual disturbance. Radiographic findings demonstrate the classic ground-glass appearance on standard x-ray films, while on computed tomographic (CT) scans there is a granular core with an occasional cystic formation, with the surrounding bony cortex generally intact.

Because of the indolent natural history of most fibrous dysplastic lesions, many of these have been traditionally observed until after puberty when the patient’s growth is complete, the risk of surgical complications is lower, and the extent of disease is defined. However, when aggressive lesions invade the orbit or orbital apex, the clinician is presented with a difficult decision regarding the timing and extent of surgical extirpation, as well as with the subsequent reconstructive requirements. Unless close observation of these lesions is performed, including detailed serial ophthalmologic examinations and serial CT scans, growth can lead to significant aesthetic and functional disabilities. The literature suggests that there may be growth after puberty in some patients, contrasting with previous statements in the literature.

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PATIENTS AND METHODS

This series involves cases limited to the maxilla and periorbital region, treated at 2 tertiary care institutions—University of Iowa Hospital and Clinics, Iowa City, and Johns Hopkins Medical Institutions, Baltimore, Md—between 1988 and 1997. At the time of treatment the patient ages ranged from 2 to 38 years. Fourteen patients with such midfacial or orbital fibrous dysplasias were retrospectively reviewed, but 6 patients were excluded who required no surgical intervention, minimal shaving for cosmetic purposes, and/or surgery for sinus disease alone. The remaining 8 patients were included in the review. Their minimum follow-up was 15 months (follow-up range, 18 months to 9 years). These patients had clinically notable orbital, nasal, and/or dental impingement by the tumor, as demonstrated by cosmetic deformity with or without functional impairment.

The preoperative and postoperative evaluation included the following physical examination findings: cosmetic deformities of the midfacial and periorbital regions, ophthalmologic evaluation, nasal dysfunction, and dental dysfunction. Radiographic evaluation, consisting of CT examinations, were obtained preoperatively and postoperatively, and in most patients CT scans were obtained to follow up any persistence or regrowth of the lesion. Table 1 reviews the presentation and procedure for each patient. Four patients will be described to demonstrate the spectrum of disease treated in our series.

This study presents a series of clinically aggressive midfacial and periorbital lesions that demonstrate the spectrum of management options. Treatment objectives emphasize preservation of important structures and function, while complete extirpation of the disease is a secondary consideration. The cases range from bulky lesions that cause primarily cosmetic and dental deformities to those with aggressively debilitating lesions of the orbit and orbital apex that require major skull base surgery and reconstruction.

REPORT OF CASES

CASE 4

A 35-year-old man had a long history of left facial pressure, symptoms consistent with sinusitis, and left-sided upper facial fullness (Figure 1). Computed tomographic evaluation disclosed an ethmoid lesion that extended into the posterior orbit and skull base and also had a cystic component in the upper maxilla (Figure 1, C). He underwent resection of the cystic component along with endoscopic drainage. As there was a resultant 4 × 3-cm defect of the medial superior maxilla and orbital rim, he underwent primary bone grafting of the orbital rim. At 15 months' follow-up, the patient's CT scan shows no abnormalities with persistence of the bone grafts; the patient has an improved appearance, and sinusitis has not recurred.

CASE 5

A 1 ½-year-old boy was seen after his parents had noted as early as 6 months of age a right midfacial mass (Figure 2). Physical examination revealed extension of the mass submucosally, intranasally, onto the alveolar ridge, and into the medial orbital floor region. A CT scan was suggestive of a solid fibro-osseous lesion of the maxilla (Figure 2, C). Because of the rapid growth of this lesion and the concern for extension into the internal orbit toward the orbital apex, it was believed that the goal of the procedure should be to remove portions of the lesion that extended into these regions (Figure 2, D). Also a concern was the apparent rapid extension toward the anterior skull base. Accordingly, through a midfacial degloving approach, the patient underwent anterior shaving of the lesion as well as a medial maxillectomy including removal of the anterior two thirds of the medial orbital floor. The latter included that portion of the ethmoid and nasal tumor that appeared to be extending toward the skull base. Postoperatively, he has continued to demonstrate growth of the lesion and has required 2 subsequent debulking procedures. While there has been growth disturbance in the right midfacial region and poor development of secondary dentition, no evidence of further extension into the orbit or skull base has been noted after 6 years.

CASE 7

At the age of 13 years, this man had right-sided epistaxis. He was apparently noted to have a right facial mass that was believed to be consistent with fibrous dysplasia, but no intervention was recommended. Over the next few years he eventually lost vision in his right eye. At age 18 years, he was referred to one of us (J.L.F.) for evaluation of his condition. His physical examination disclosed a severely deformed midface bilaterally, with a mass completely obliterating the right nasal cavity, and a friable mass filling the palate and two thirds of the maxillary antrum. There were severe dental deformities and evidence of bleeding from the gingiva around the distorted teeth (Figure 3). He also had a nonfunctioning, distorted, blind right eye. Computed tomographic evaluation confirmed a large centrally necrotic mass of the right maxilla, crossing the midline into the left maxilla and obliterating the right nasal cavity. There was extension into the skull base and orbital apex, and this was believed to be the cause of his blindness at an earlier age. There was obliteration of the palate and the alveolar ridge. As the skull base component to this lesion did not appear to involve the contralateral orbital apex region, it was believed that a complete skull base resection would not be required but could be followed up by subsequent serial CT scans, which had not shown change over 1 year.

Accordingly, through a midfacial degloving approach, this patient underwent total bilateral maxillectomy, leaving only the pterygoid plates intact (Figure 3, D). He also had...
During this resection, the patient went into disseminated intravascular coagulopathy necessitating cessation of the procedure. After a difficult postoperative course and correction of his coagulopathy, the patient secondarily underwent reconstruction of his osseous skeletal defect with a scapular free flap with a soft tissue paddle (Figure 3, D). He subsequently has done well without evidence of tumor regrowth and has undergone functional recon-
struction of his maxilla with osseointegrated implants with subsequent dental prosthetic placement.

CASE 8

A 6-year-old girl had a history of a rapidly growing right midfacial mass with orbital deformity (Figure 4). On physical examination she was noted to have a proptotic and laterally positioned globe and extensive right facial mass. Computed tomographic scan confirmed a fibrotic and centrally necrotic lesion of the maxilla with extension into the ethmoids and anterior skull base (Figure 4, C). Extension into the orbital floor and medial wall was significant. Because of the history of rapid growth, impingement on the right orbital cavity as well as encroachment on the orbital apex and the skull base, it was believed that the patient should undergo a partial resection of the maxillary component of this lesion including a medial maxillectomy, as well as an anterior craniofacial resection of the expanding cribiform component and right superior medial orbital aspect of the lesion. This also included the entire orbital floor and medial wall. This was accomplished through combined midfacial degloving and coronal approaches (Figure 4, D). A frontal craniotomy and a frontal bar osteotomy provided access to the anterior fossa component of the lesion. Calvarial and rib bone grafts were used to reconstruct the orbital rim, orbital floor, and medial wall, while a pericranial flap was used to separate the intercranial and sinonasal cavities.

Postoperatively the patient was noted to have no vision in the ipsilateral eye after not being awakened until 24 hours postoperatively. Computed tomographic scan at that time showed that the orbital floor bone graft was close to the inferior aspect of the optic canal. There was concern about returning this patient to the operating department, and as it was more than 24 hours after surgery, the combined surgical team of otolaryngology, oculoplastic surgery, and neurosurgery elected to treat her with high-dose steroids. She has done well with no reoccurrence of her lesion. Eight years postoperatively she has some evidence of midfacial growth restriction and a persistent mild globe malposition (Figure 4, D).

RESULTS

The results are given in Table 2. Postoperative evaluation was performed and involved physical examination findings, comparisons of preoperative and postoperative photographs, and, in most cases, CT scan evaluation. The range of follow-up was 18 months to 9 years, with 5 of the 8 patients being followed up for 4 years or longer. Cosmetic appearance was improved in all 8 patients, with the most substantial improvement seen in those with the worst deformities. All patients with a maxillary or midfacial deformity had significant improvement in appearance. Patient 8 had severe exophthalmos and globe malposition from her lesion. Postoperatively, she had significant improvement of
the exophthalmos although there was moderate persistence of globe malposition along with the unfortunate iatrogenic blindness. Patient 7 who had preoperative blindness had no significant change in his orbital deformity as no resection was performed in the periorbital region. All other patients had mild preoperative orbital deformities; these patients had subtle improvements of this malposition. Of the 3 patients who had maxillary fibrous dysplasias during the period of primary dentition, 1 proceeded to develop normal secondary dentition, 1 had limited eruption of secondary dentition, and it is too early in the last patient's follow-up period to determine whether secondary dentition will occur.

Regarding follow-up on reconstruction, of the 3 patients who had free bone grafts, 2 have demonstrated complete persistence of these bone grafts while 1 has shown partial resorption that has not affected the physical appearance or further growth. In the patient with the osseous free flap, this neomaxilla is stable at 5 years after reconstruction and is functioning normally.

Regarding fibrous dysplasia regrowth, there is evidence of regrowth in patients 5 and 6. Patient 5 has required subsequent limited resections in his 6 years of follow-up. This patient also has significant growth disturbance of the maxilla and may require subsequent reconstruction and/or orthognathic surgery. Only 18 months after resection, patient 6 who also had evidence of regrowth at the time of his last examination had not yet demonstrated a need for further resection despite involvement of the skull base. Finally, patient 1 has not demonstrated regrowth but has a significant dentofacial deformity including malocclusion and a maxillary shift with an elongated maxilla. He is being considered for orthodontics and orthognathic surgery.

**COMMENT**

The management of midfacial and periorbital fibrous dysplasia is generally conservative, with many patients not requiring any surgical intervention unless significant cosmetic deformities or functional problems exist, such as chronic sinusitis nasal obstruction, visual disturbances, or dental problems. A small subset of patients exists who have significant cosmetic deformities such as midfacial and cheek protrusion (the so-called lion's face) or globe malposition, with or without the addition of functional problems. Globe malposition becomes particularly important when there is impingement on the orbit contents such that vision is affected (diplopia or visual acu-
ity changes), progression of nasal obstruction, or impingement on the dental alveolus. While cosmetic deformities generally occur before functional problems, the combination of both occurs in severe cases. These issues are important in all age groups but are particularly problematic for children. Lesions arising in younger children, while facial and dental growth are occurring, present a significant therapeutic dilemma.

The literature is replete with opinions on how to best manage each of these functional and aesthetic components. However, there is disagreement as to the acceptance of the common opinion that these fibro-osseous lesions stop growing after puberty when the patient’s growth is complete. Unfortunately, it was the belief in the premise of growth cessation by physicians managing patient 7 that led to his blindness by disease progression. This patient progressively lost vision in his midteens and was blind by the time he was first evaluated by the treating surgeons at age 17 years.

Accordingly, we believe that the evaluation and treatment must be tailored to each patient considering the following issues: age of presentation, the presence of documented growth, direction of growth and impingement on vital functional structures, whether the lesion has a cystic component, and whether there is cosmetic deformity.

The evaluation of patients with possible fibrous dysplasia involves radiographic diagnosis. Our current standard is axial and coronal CT evaluation that will generally reveal the granular fibrous bony appearance typical of fibrous dysplasia. This scan is also important to determine if there is any cystic degeneration of the lesion. Such cystic lesions were noted in 3 (patients 4, 6, and 7) of our 8 patients and were important findings for eventual reconstructive demands. Pathologic diagnosis is ideal although it can generally be obtained at the time of partial surgical resection. In lesions including the nasal cavity, transnasal endoscopic biopsy may be useful. The history of growth as described by the patient or by the patient’s family is extremely important. If there is rapid growth by history, particularly in a child, we would be more inclined to intervene surgically at an earlier stage, particularly if there is evidence on CT scan of invasion of important structures such as the orbit, anterior skull base, and nasal cavity. Invasion into the inferior maxilla presents a difficult challenge because of the potential involvement with the dentition. This is amplified further in children who have not had eruption of secondary dentition.

In the patient who has a lesion that has been determined to be stable or limited in growth and without impingement on important functional structures, observation with serial CT scans is indicated and acceptable. Serial ophthalmologic examinations are important in all peri-orbital lesions. However, when there is evidence of rapid growth by history and impingement of important
structures by CT scan, surgical intervention is recommended. The decision at this point becomes the nature of the resection. Some authors recommend complete extirpation of upper midfacial and orbital lesions whether or not there is evidence of continued growth. However, we perform complete resection of such periorbital lesions only when there is evidence of persistent dangerous growth into the posterior orbit and skull base. Otherwise, our recommendation includes decompression of the orbit and restoration to the normal orbital shape to allow for return of globe position. Similarly, particularly in the child with primary dentition, the resection of the maxillary component is generally conservative and restricted to contouring of the anterior maxilla for cosmetic purposes, possible limited medial maxillectomy for functional purposes, and occasional shaving of excessive impingement of the fibrous lesion into the gingival areas. The latter may cause both a cosmetic deformity as well as create a functional problem over the gingival area.

Figure 5 and Figure 6 offer algorithms for treatment and reconstruction of orbitomaxillary fibrous dysplasia. Figure 5 suggests our recommendation for management of the cosmetic deformity and Figure 6, the management of functional issues involving the orbit, nose and sinus, and dental alveolus.

Counseling is very important in the preoperative stage for outlining the advantages and disadvantages of the degree of resection. The patient and family must understand that resection of the tumor may be incomplete, and because in many cases complete resection would generally require removal of the alveolus (and dentition), resection of the lower maxilla is avoided except in extreme circumstances (as in case 7). Similarly, resection of a significant portion of the skull base is fraught with potential complications that may outweigh the benefits. They must also be aware of the slight risk for malignant degeneration of these tumors and be allowed to weigh the advantages and disadvantages of aggressive vs conservative resections.

All 8 patients in our series were approached through a midfacial degloving approach that allowed for adequate exposure for resection, and when necessary, reconstruction. While only patient 8 in our series had a combined coronal craniofacial approach along with the midfacial degloving, this is an excellent combination of approaches for difficult lesions of the upper midface and skull base. As no facial scars are created, this is particularly useful in the pediatric population although we recommend it in adults as well. Through these approaches, the resection was then tailored for each patient depending on their cosmetic deformity and functional symptoms as well as whether there was evidence of active growth. As 2 of our younger patients (patients 5 and 6) had evidence of aggressive active growth at the time of diagnosis, resections were more aggressive with the basic premise being to resect the lesion where we believed it could do the most harm if left untreated. Accordingly, orbital and ethmoid resections along with the upper medial maxilla were performed in these patients. However, resection of the lower maxilla was limited and the dentition was undisturbed. Both patients have demonstrated regrowth, one of whom had undergone subsequent conservative debulking procedures. The other patient will likely require further resection in the future.

Three of our patients had cosmetic deformity and mild globe malposition. In these patients, anterior maxillary debulking with limited orbital floor resection was performed. All of these young adult patients have done well and have not required further resection of their lesions.

Table 2. Deformity, Follow-up, Regrowth, and Dental Restoration Status

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Deformity Status</th>
<th>Duration of Follow-up, y</th>
<th>Recurrent Growth*</th>
<th>Dental Restoration Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Limited orbital, moderate midfacial, malocclusion, and elongated maxilla</td>
<td>No orbital, limited midfacial, malocclusion, and elongated maxilla</td>
<td>4</td>
<td>No</td>
</tr>
<tr>
<td>2</td>
<td>Limited orbital and moderate midfacial</td>
<td>No orbital and minimal midfacial</td>
<td>6</td>
<td>No</td>
</tr>
<tr>
<td>3</td>
<td>Moderate midfacial and zygomatic</td>
<td>Limited midfacial, and zygomatic</td>
<td>2</td>
<td>No</td>
</tr>
<tr>
<td>4</td>
<td>Moderate lateral nasal, infraorbital, and superior midfacial</td>
<td>Minimal deformity</td>
<td>1.3</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>Severe midfacial</td>
<td>Limited midfacial, and disrupted secondary dentition</td>
<td>6</td>
<td>Yes</td>
</tr>
<tr>
<td>6</td>
<td>Moderate orbital and moderate midfacial</td>
<td>Limited orbital and limited midfacial</td>
<td>1.3</td>
<td>Yes</td>
</tr>
<tr>
<td>7</td>
<td>Severe orbital (including blindness), severe midfacial, dental, and palatal (necrosis)</td>
<td>Blindness, minimal, midfacial, and prosthetic dental (functioning)</td>
<td>5</td>
<td>No</td>
</tr>
<tr>
<td>8</td>
<td>Severe exophthalmos and midfacial</td>
<td>Moderate orbital, minimal midfacial, and blind right eye</td>
<td>8</td>
<td>No</td>
</tr>
</tbody>
</table>

* Only patient 5 required 2 surgical resections because of recurrent growth.
Four of the 8 patients required no form of reconstruction. We believe that the defects left by the partial orbitectomies in several other patients did not require reconstruction as they were performed for decompression alone. However, based on radiographic evidence, 4 of our patients were diagnosed as having cystic lesions, an important issue to consider when performing even limited resection of a lesion. In this situation, a cystic cavity will be entered on dissection thus creating a defect where there was once excessive protrusion of tissue. Accordingly, we were prepared in each of these 4 cases to perform primary reconstruction of the cystic defect using free calvarial bone grafts.

The 2 patients with the largest and most debilitating lesions required careful preoperative planning, both in terms of the limits of the resection and for subsequent primary reconstruction. The 6-year-old patient (case 8) who had grossly evolving proptosis and evidence of progressive skull base growth required a combined craniofacial approach for subtotal resection of the lesion. She underwent reconstruction of her orbital rim and upper maxilla with free bone grafts, as well as the use of similar grafts for reconstruction of her entire floor and medial wall of the orbit. This patient was the first case of our series; surgery was performed in 1988. Since then, orbital reconstruction patients are either awakened immediately for visual examination or have immediate postoperative CT scans to confirm safe position of the reconstruction material in the posterior orbit. She has proceeded with adequate facial growth with a satisfactory cosmetic appearance and has had no evidence of lesion regrowth, while her regrettable loss of vision has not limited her development. The other significantly challenging patient (patient 7) was the 20-year-old man who had the grossly deforming necrotic maxillary lesion that had, at an earlier stage, caused blindness in the ipsilateral eye because of unabated growth into the skull base. By the time of our evaluation, serial CT scans suggested no further regrowth into the orbital apex and skull base so it was decided that this portion of the lesion could be left undisturbed. However, the lesson to be learned is that had he been adequately evaluated at an earlier stage, an aggressive decompression at the orbital apex would certainly have been indicated. Regardless, because of the progressive necrosis of his lesion and the subsequent loss of functional dentition, our approach involved a total bilateral maxillectomy to resect completely the necrotic portion of his lesion. This was followed up by bony reconstruction of an adequate nature to allow for secondary placement of dental implants and dental rehabilitation. Both of these cases are unusual and have represented the most significant resection and reconstruction challenges in our series.

CONCLUSIONS

We believe that a tailored approach should be undertaken in the evaluation and resection of patients with aggressive midfacial and periorbital fibrous dysplasia. The degree of resection will vary according to the aggressiveness of the lesion, the age of the patient, and the nature of involvement of surrounding functional structures. Complete resection of the lesion is rarely necessary. Similarly, while reconstruction is only occasionally required, preoperative planning should include a precise evaluation of the degree of resection to allow for adequate planning of reconstructive needs. Finally, coun-
Counseling is important in all fibrous dysplasia cases with respect to a review of vital structure involvement and reconstructive requirements. Potential growth restrictions in children caused both by the presence of the lesion and by any surgery that is performed should be reviewed. Similarly, families with young children who have these lesions must be made aware that secondary dentition may not erupt normally, and they may require secondary orthodontics, prosthodontic restoration, and even orthognathic surgery.

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