Fibrous Dysplasia of the Maxillary Bone
A Case Presentation

Author: Kevin T. Kavanagh, MD, MS, FACS. Clinical Associate Professor, University of Kentucky, Department of Surgery, Division of Otolaryngology. Somerset, KY, USA.

Abstract: A case of monostotic fibrous dysplasia in a 16 year old boy is presented. A review of the literature and treatment options for the patient are discussed along with flash presentations of the patient's computed tomographic scan, three dimensional reconstructions and video of operative shaving of the patient's maxillary bone.

Introduction: Fibrous dysplasia is an idiopathic disease which causes progressive expansion and deformity of bones. Histologically, it is a fibrous osseous lesion where fibrous tissue replaces laminar bone and the bony component is haphazardly organized. Fibrous dysplasia can involve multiple bones (polyostotic) or a single bone (monostotic). In 10% of monostotic fibrous dysplasia and 50% to 100% of polyostotic fibrous dysplasia, there is involvement of the facial and cranial bones.¹

Males are affected as commonly as females¹ and usually first presents before the third decade of life² A more aggressive form of the disease, McCune-Albright syndrome, is associated with a triad of café-au-lait spots, precocious puberty and fibrous dysplasia.³ ⁴ ⁵ Fibrous dysplasia usually has a relatively benign course with the major symptom being cosmetic deformity, which may become severe. Malignant transformation has been reported in up to 0.5% of patients.⁶ However, vital structures such as the visual apparatus may also be affected with resultant loss of function. When this happens surgical intervention is indicated.

This report will present a case of isolated fibrous dysplasia of the maxilla in a 16 year old male.

Case Report: The patient was a 16 year old male who had fibrous dysplasia, first diagnosed at eight years of age. At that time, the patient underwent a biopsy to rule out a sarcoma. Spongy fibrous tissue was found at the time of biopsy. Note the expansion of the cheek with the elevation of the eye and depression of the lip.
A three dimensional (3-D) reconstruction of the patient’s computed tomographic (CT) scan. Note the expansion of the right maxillary bone with the inferior displacement of the teeth and alveolar ridge and the superior displacement of the orbit. The infraorbital rim is also displaced forward. The FLASH animation below shows the 3-D CT scan and the upper right-hand button will play a video of the surgery.

At 19 days post operatively, the patient was pleased with the result. Despite extensive surgery, there was still significant asymmetry but less than the preoperative appearance. The patient reported only minor numbness in the midcheek. Further reduction will probably be necessary in the future, but extensive surgery should wait until the patient is in the mid-twenties.  

Discussion: Clinical symptoms arise from the expansion of the bone which compresses adjacent structures. The disease is progressive but with time will often burn out and stabilize. Fibrous dysplasia may stabilize after puberty. However, the disease may progress into adulthood and observing the patient for several years before cosmetic surgery is undertaken is prudent. Even if this is done, progression in later life may occur. The type of surgery performed varies from shaving and contouring of the bone to radical surgery. Facial contouring is the least invasive.
but may need to be performed repeatedly due to suboptimal results and recurrence. Complete resection may be required in patients with Albright Syndrome since there is a high incidence of progressive growth and recurrence. For large cosmetic deformities, recontouring and repositioning of bone may be required. 3-D CT scan reconstruction of the patient’s facial bone can help in surgical planning.

Orbital involvement with loss of vision is one of the most severe but relatively uncommon complications of fibrous dysplasia. Surgical intervention is indicated for visual impairment caused by fibrous dysplasia but prophylactic decompression is controversial.

Medical therapy with bisphosphonates may stabilize the lesions and play a role in prevention of progression of the disease in adults. Radiation therapy is contraindicated because of a reported incidence of malignant degeneration with the formation of osteosarcoma.

The reported patient, had no progression of the disease for three years, at this time the patient underwent facial contouring. The operation was performed through the gingival-buccal sulcus (see left-hand picture). The facial tissues and periosteum were elevated off the maxillary bone. Hard ossified bone was encountered which was slowly removed using a drill. This was in contrast to the tissue found eight years previously at the time of biopsy which was dark and grainy and easily removed with a curette. Care was exercised to avoid injury to the infraorbital nerve (see right-hand picture).

The patient and family felt that the contouring of the patient’s maxilla resulted in a significant improvement of his cosmetic appearance and improvement in the appearance of his alveolar ridge. The outside observer may view the improvement as mild to moderate. This case illustrates the debate between major and minor surgery for fibrous dysplasia. The inferior approach through a buccal incision gave a restricted view of the area of the infraorbital rim and almost no access to the zygomatic arch. Improved access could be obtained by cutting the infraorbital nerve but this would result in a numb cheek. The infraorbital rim and zygomatic arch are not only enlarged but displaced laterally and tent the tissues of the cheek forward. Thus, to obtain a major improvement, an additional incision below the eye lash line (subciliary) with extensive recontouring and osteotomies with resetting of the bone would be required. This is illustrated by the case reported by Gosain, whose patient underwent two bone shavings before undergoing major surgery. However, major surgery should be done with caution in a young patient where recurrence and progression may occur which would require further surgery to maintain his cosmetic appearance. The best recommendation is to follow the wishes of the patient.

**Conclusion:** Surgical intervention in patient's with fibrous dysplasia should be reserved to correct functional impairments or significant cosmetic deformities. Patients will have to weigh the degree of surgical intervention they wish to endure versus the expected cosmetic improvement. Although, the disease often burns out after puberty, it may become progressive or recur in early adulthood. This is more common in Albright's Syndrome. In these patients, surgical resection of the entire lesion should be considered.
References:


