CASE REPORT

Prophylactic optic nerve decompression in pediatric craniofacial fibrous dysplasia: To do or not to do?

R Keerthi, Tulasi Nayak

Department of Oral and Maxillofacial Surgery, Vokkaligara Sangha Dental College and Hospital, V. V. Puram, Bengaluru, Karnataka, India

Abstract

Fibrous dysplasia (FD) is a pathological condition in which normal bone is replaced by abnormal fibro-connective tissue. Although characterized as a benign condition, it can be large and invasive, causing considerable disfigurement and dysfunction. When faced with such a destructive process in a child, the obvious questions raised are what to do about it and when. We present here a case of craniofacial FD in an 11-year-old boy, in whom the growth in the right orbit had caused swelling, epiphora, proptosis, and pain. The patient was treated with prophylactic optic nerve decompression with excellent results. While deteriorating vision is a definitive indication for optic nerve decompression, the need for a prophylactic treatment is a controversial one. The rate of irreparable loss of vision is well-documented, and the resulting morbidity is one which must be considered in children. We have singularized this case of prophylactic optic nerve to review the various clinical and treatment aspects of craniofacial FD and to specifically debate the viability of prophylactic optic nerve decompression in pediatric craniofacial FD.

Keywords

Craniofacial, decompression, optic nerve, pediatric fibrous dysplasia, prophylactic

Introduction

Fibrous dysplasia (FD) is a disease of bone maturation and remodeling in which the normal medullary bone and cortices are replaced by a disorganized fibrous woven bone.[1] The combination of a lack of stress alignment and insufficient mineralization yields substantial loss of mechanical strength.[2] While FD can occur in any bone of the body, the jaw bones, ribs, femur and tibia are most commonly affected.[1,2] Among the jaw bones, the body of the mandible and posterior tooth bearing regions of the maxilla are most commonly affected.[1]

Although FD becomes clinically apparent between the ages of 5 and 15, it begins as spontaneous mutation or deletion of a bone maturation transducer protein in the embryo. All the daughter cells of this mutated cell line produce the disorganized bone.[1] When it involves the jaw-midface-skull complex, it is termed as craniofacial FD.[1] This type of FD presents with a significant aesthetic and functional impact. The treatment modality is controversial, ranging from medical to surgical which in turn may be conservative or radical. The preferred approach to craniofacial FD is no treatment.[1] However, progressive deformities, pathological fracture, and bone pain are inevitable in skeletally immature patients as the lesions tend to enlarge over time.[1] These symptoms or a psychological need may necessitate surgical intervention.

We, in our case, have visited the need for prophylactic decompression of the optic nerve as a mainstay for patients who present with any orbital symptom which necessitates surgical intervention.

Case Report

An 11-year-old boy submitted to the Department of Oral and Maxillofacial Surgery with his parents in February 2014, with 6 months history of a swelling in the right middle half of the face, pain in the right globe and associated epiphora. The parents reported that the appearance of the boy’s right face was asymmetrical compared to the left side, and the deformity had gradually progressed during the past 6 months. No notable pain was present on palpation. The patient had a normal physical development and no history of trauma. The patient did not present with visual disturbance, sinusitis, tinnitus, or paresthesia. The physical examination revealed a bony enlargement extending from the right frontozygomatic region to the right corner of the mouth with no raise in local temperature.

Intraorally the bony enlargement was present over the right half of the maxilla extending into the vestibule and palate opposite the lateral incisor to the first molar. The patient was examined...
for pigmented lesions and signs of precocious puberty which were absent. On ophthalmic examination, no abnormalities were noted in the vision or extraocular motion; no exophthalmos or proptosis was present. The history and clinical findings indicated a fibro-osseous lesion. The differential diagnosis ranged from FD, osteosarcoma, ossifying fibroma, maxillary sinus osteoma, fibrous sarcoma, aneurysmal bone cyst, giant cell tumor, odontogenic tumor, ossifying fibroma, osteochondroma, osteoblastoma, and cementifying fibroma.

A computed tomography (CT) of his craniomaxillofacial region showed a large expansile lytic lesion with a “ground-glass” appearance in the right squamous and petrous part of temporal bone, right zygoma, right medial and lateral pterygoid plates, right wing of the sphenoid, clinoth process, right maxillary, sphenoid and ethmoid sinuses, lateral wall of the right orbit, right alveolar process of the maxilla, right tympanic cavity, and symphysis menti. The margins of the lesion were ill defined. The cortex showed a marked expansion and no periosteal reaction or distinct soft tissue mass [Figure 1a and 2a]. The mass within the orbital cavity showed a compression of the globe [Figure 1a]. Based on the clinical and radiographic features, a provisional diagnosis of polyostotic FD was made. A full body scintigraphy (Tc99m) revealed a hot focus on only the right craniomaxillofacial region, blood urea was within normal limits (15 g/dl) and creatinine was below normal limits (0.5 mg/dl), alkaline phosphatase levels were high (309 IU/l) indicating an increased turnover of bone. The parathormone levels were found to be subnormal (7.66 pg/dl) with normal calcium (9.9 mg/dl) and normal phosphorous levels (4.5 mg/dl).

A bone biopsy was performed at the right maxillary vestibular region. Histological examination showed a regular parallel arrangement of trabeculae of immature bone in a fibrousvascular stroma with focal areas showing curvilinear eosinophilic trabeculae comprising of osteocytes in lacunae. No osteoblastic rimming was seen and focal areas of hemorrhage had been noted. The parallel arrangement of the trabeculae suggested the lesion was mature, and the hypocellular stroma differentiated the lesion from ossifying fibroma.

The overall clinical, radiographic, and histological features were suggestive of polyostotic FD of the craniofacial region without McCune–Albright syndrome.

A multidisciplinary meet between the neurosurgeon, ENT surgeon, Ophthalmologist, and Maxillofacial surgeon found that a radical excision of the abnormal tissue was not feasible. However, the presence of the symptoms due to bony enlargement ascertained the need for immediate treatment. The team hence opined that a conservative surgery to decompress the globe and optic nerve would be appropriate.

A conservative surgery encompassing debulking and recontouring of dysplastic bone was performed at the lateral wall of the orbit, zygomatic arch, and maxillary alveolus under general anesthesia to relieve the globe and recontour the area. An upper eyelid incision was used to approach the dysplastic tissue. A subperiosteal dissection was carried out to expose and recontour the dysplastic tissue within the orbit, the frontozygomatic region, and the malar prominence. Care was taken to preserve the attachment of the lateral canthal tendon, optic nerve, the globe, the contents of the infraorbital canal, and the lacrimal gland. An intraoral vestibular incision was used to expose and recontour the dysplastic tissue on the maxillary alveolus and zygomatic buttress regions [Figure 1b and 2b]. No post-operative complications have been present, and the patient has been asymptomatic since. The patient’s parents were informed of a very high possibility of recurrence and have hence been kept on a regular follow-up regimen. The follow ups consist of a regular assessment of ophthalmic function and radiographic study.

**Discussion**

**Etiology and pathogenesis**

First described by von Recklinghausen[4] in 1891, it was Lichtenstein in 1938 who recognized the condition as a distinct entity and suggested the name “FD.”[5] It accounts for about 2.5% of all bone lesions and 7% of all benign bone tumors.[6] It presents in childhood or early adolescence, typically arresting at puberty.[7] The disease may involve a single bone (monostotic) or multiple bones (polyostotic), and it may present in association with McCune–Albright syndrome[8,9] or Mazabraud’s
syndrome.\textsuperscript{[10]} Jaffe-Lichtenstein syndrome, describes polyostotic FD with cutaneous melanotic pigmentation in the absence of endocrine abnormalities.\textsuperscript{[1]} Although a genetic disease, the lack of vertical transmission, and the observation that the lesions tend to lateralize and respect the midline has helped to establish that the disease is a result of postzygotic mutations, and thus, the patients are somatic mosaics.\textsuperscript{[11-13]} The etiology has strongly been linked with a mutation in the Gsa gene that is located at chromosome 20q13.2-13.3.\textsuperscript{[14]} This mutation was first noted in patients with McCune–Albright syndrome and later demonstrated in patients suffering from monostotic and polyostotic FD.\textsuperscript{[14]}

Bianco et al., Bianco and Robey\textsuperscript{[22,25]} first demonstrated the role of the bone marrow stromal cell (BMSC) in FD. They showed that BMSCs differentiate along the osteogenic lineage, but the activating mutations in the GNAS gene arrest the differentiation, hence giving rise to the fibro-osseous tissue [Table 1].

In >95% of the cases of FD, arginine is replaced by either cysteine or histidine (R201C or R201H) or more rarely other amino acids.\textsuperscript{[18,19]} These mutations inhibit the intrinsic GTPase that leads to the formation of a ligand-independent intracellular cAMP. This defect also explains the manifestations of the cafe-au-lait spots, precocious puberty, and hyperthyroidism.\textsuperscript{[18]} The extent and type of FD are determined by the timing of the mutation.\textsuperscript{[20]} An earlier mutation leads to a wider distribution of the disease where the mesoderm, ectoderm, and endoderm, i.e., the bone, skin, and endocrine system are all involved. For disease that is isolated to the skeleton but involves craniofacial and long bones, the mutation must have arisen later in development in the meso ectoderm, as the craniofacial bones are of a neuroectodermal origin, and long bones of mesodermal origin.\textsuperscript{[18]}

Other suggested etiologies include trauma to bone with an underlying developmental defect or a hamartoma-like overgrowth of fibrous tissue. However, neither of the suggestions could explain endocrine changes and pigmentation seen in some of the polyostotic cases; for these phenomena some congenital abnormality in the hypothalamus was thought to be responsible.\textsuperscript{[21]} Another hypothesis is that the increased number of osteoclasts and subsequent bone resorption seen in FD are due to elevated interleukin 6 levels. This hypothesis has been supported by the reduction of spread of FD into the normal adjacent bone using pamidronate (a potent inhibitor of osteoclasts).\textsuperscript{[21]}

Clinical picture
Craniomaxillofacial FD refers to the involvement of multiple bones in the craniofacial skeleton with 90% of the cases presenting before the age of 5 years\textsuperscript{[20]} and >90% of the lesions established at all sites before the age of 15 years.\textsuperscript{[12]} These lesions tend to gradually becomes quiescent and finally with the termination of skeletal growth becomes inactive and ceases to grow.\textsuperscript{[21]} There is however some report which has shown continued growth in adults.\textsuperscript{[21]} The craniofacial bones are affected in about 10% of cases of monostotic FD and in 50% to 100% of cases of polyostotic FD.\textsuperscript{[21]} Lustig et al.,\textsuperscript{[24]} have mentioned that the most affected craniofacial bones are the ethmoid (71%), sphenoid (43%), frontal (33%), maxillary bones (29%), and less frequently the temporal (24%) and occipital bones (5%). While other studies have reported that the frontal bones were most commonly involved followed by the sphenoid, ethmoid, parietal, temporal, and occipital bones.\textsuperscript{[22]} FD of the jaws affects the maxilla more frequently than the mandible.\textsuperscript{[21]} It is interesting to note that when the vault of the skull is affected, the mass grows mostly outward, but when the base is affected, the mass grows into the cranial and/or orbital cavities, producing the usual signs of space occupying lesions.\textsuperscript{[21]}

Even though there have been considerable variations among authors regarding the epidemiological data of craniofacial FD, A meta-analysis conducted by Wu et al.,\textsuperscript{[20]} in 2014, has shown that the monostotic and polyostotic forms of FD occurred at similar rates with unilateral FD being more common than bilateral form. They have also found no significant difference between the prevalence of male and female FD cases.

FD most presents as a slow growing lesion with an indolent course, subsequently leading to facial deformity and compression of adjacent vital structures such as optic nerve, the facial nerve, the globe, the nasal cavity, and the middle ear ossicles.\textsuperscript{[25]} The patients may present with pain, paresthesia, facial asymmetry, deteriorating vision, hearing impairment, and nasal congestion. Quite often, the patients may also present with malocclusion.\textsuperscript{[27]} The explanation for the bony pain is not clear nor is there any obvious association between the expanse of the disease and the pain experienced; with some patients with limited bone involvement experiencing excruciating pain while others with extensive disease may be relatively pain-free, and vice-versa.\textsuperscript{[18]}

The intraoral changes occur gradually.\textsuperscript{[22]} Akintoye et al.\textsuperscript{[28]} examined 32 patients with craniofacial FD and found that 28% of the patients had dental anomalies within FD bone. The most recurrent anomalies included: Oligodontia, retained deciduous teeth, tooth rotations and displacement, taurodontism, and enamel hypoplasia. About 10 of these patients had received orthodontic therapy to correct malocclusions. However, the duration was longer than the conventional treatment (approximately 2-4 years), with less than satisfactory results and a higher tendency of relapse.

Histological features
Histologically, FD usually exhibits he classically described “Chinese characters.” However in FD, the degree of activity of the lesion determines its microscopic features. In active cases, the histological picture is composed of highly cellular
connective tissue matrix, containing numerous dark staining fusiform and stellate cells with many mitotic figures and the absence of intracellular collagen. Over time, FM may show maturation, which is characterized by the formation of lamellar bone and parallel arrangement of the trabeculae as seen in our case. Histologic features alone, however, must be correlated with a clinical and radiographic presentation for a definitive diagnosis.

**Radiographic features**

Historically, the radiographic presentation of FD is described as ground glass in panoramic radiographs and as orange peel in intraoral periapical dental radiographs(Iseri in his review mentioned that Fries (1957) classified the radiological appearances of FD of the craniofacial region into three types: Pagetoid (56%), sclerotic (23%), and cyst-like (21%). In a study conducted by Akintoye et al., the ground glass pattern was most prevalent in the maxilla and mandible of patients below 21 years, as seen in our patient; but in patients over 21 years, the radiopaque pattern was most prevalent in the maxilla while mixed radiolucent/radio-opaque pattern was most prevalent in the mandible. No radiolucent FD lesions were observed in the maxilla compared with 12.5% of cases observed in the mandible. Based on the radiographic features, FD in children has been divided into an extended type and a circumscribed type by Andrisano et al. Circumscribed variety describes the involvement of less than a quarter of the involved bone and only one cortex, and the extended type is one where more of the bone segment and both cortices are involved. CT Based on the CT findings, three patterns of FD have been described in the CT: Ground-glass pattern, homogeneously dense pattern, and cystic variety.

In general, the maturation of the disease is reflected on the CT. With aging, the lesion tends to appear "stable," i.e. characteristically bounded by a sclerotic rim and surrounded by a thick reactive bone. On the contrary, an aggressive, unstable lesion is characterized a lesion which makes the cortex thin and expansile without a distinct sclerotic rim.

**Table 2: Zones and treatment of craniofacial FD**

| Zones | Areas involved | Treatment prescribed |
|-------|----------------|----------------------|
| Zone 1 | Frontal, orbital, nasal, ethmoid, zygoma, and upper maxilla | Complete excision and reconstruction |
| Zone 2 | The parietal bone, part of the occipital area and the lateral cranial base of the temporal zone | Conservative or radical resection |
| Zone 3 | Central cranial base, petrous, mastoid, pterygoid, and sphenoid bones | Dangerous to manage, Usually nonoperatively in the asymptomatic patient |
| Zone 4a | Includes the maxillary alveolar bone | Conservative excision and recontouring |
| Zone 4b | Includes the mandible | Conservative excision and recontouring |

FD: Fibrous dysplasia

**Treatment**

The treatment of FD may include surgical or medical management of symptomatic patients. In cases unmanageable by medical management, definitive surgical treatment should be the mode of action.

Chen and Noordhoff and Ricalde and Horswell classified the skull into 4 zones and emphasized the treatment and reconstruction of each zone in craniomaxillofacial FD [Table 2].

Even though a complete resection of all the dysplastic tissue containing the mutated gene is ideal, it is often a risky endeavor with a large number of possible complications.

Due to the biological features of FD, curettage is associated with a high recurrence rate as not all of the mutated cells can be removed. Li et al. reviewed older literature and found a 100% recurrence rate when treated with emptying and curettage. Hence, the surgical removal of all dysplastic tissue is ideal to prevent recurrence, and wide excision of the involved bone has been advocated with a few successful treatments in adults. However, this treatment protocol has not been tested in pediatric cases.

Murray et al. and Adetayo et al. reported a reliable and consistent result by designing a three-dimensional surgical imaging technology to virtually simulate tumor excision and designed a polyactic-co-glycolic acid-implantable graft framework for reconstruction.

**Involvement of the orbit**

Goisis et al. have mentioned that when the orbit is involved, immediate decompression is mandatory to prevent errors in refraction, focusing, and accommodation. Deteriorating vision is an absolute indication for surgical decompression with high-dose corticosteroids being used in conjunction. Involvement of the orbit and optic nerve warrants a comprehensive neuroophthalmologic examination. Controversy, however, rages on, regarding the effectiveness of prophylactic decompression. Prophylactic decompression is based on the belief that visual loss is directly related to optic canal stenosis. Chen et al. recommend prophylactic decompression in two circumstances. First, the radiological involvement of the optic canal; the second is in regards to the speed with which visual deterioration can occur and the brief amount of time before it becomes permanent. However, the case-control study conducted by Lee et al. showed that out of 49 canals which were completely encased by FD, only 2 had a visual impairment. They hence concluded that diagnostic images alone are insufficient to determine the necessity for prophylactic decompression. Similarly, other studies have also propagated surgical intervention in cases with visual impairment. Thus, the correlation of canal stenosis and visual loss is not completely clear. In fact, it is insinuated that visual may be due to a primary or secondary mass lesion rather than optic canal stenosis.
Our patient had presented with dysplastic tissue in all the zones mentioned above [Table 2] and radical surgery was deemed too risky. The involvement of the orbit and pain in the globe with epiphora (not associated with stenosis of the nasolacrimal duct) prompted us to surgically relieve the pressure on the globe and also prophylactically decompress the canal even though the visual acuity was normal.

Medical management

Bisphosphonates are the only medications that have shown any efficacy in the treatment of FD and are pain relief is the only clear indication for its use. However, Leet and Collins have found that disease in the craniofacial region can be less responsive to bisphosphonates. Similarly, Chapurlat and Orcel have mentioned that pamidronate is less efficacious in about 15% of the patients and in some patients, there was an initial positive response followed by a relapse in bone pain or inability to maintain reduced levels of biochemical markers of bone turnover.

The safety of bisphosphonates in FD has been found to be satisfactory. However, various studies have reported safety issues with the commonly used bisphosphonates such as pamidronate, alendronate, risedronate, and zoledronic acid. Short-term safety issues such as hypocalcemia, atrial fibrillation, ocular toxicity, renal toxicity, gastrointestinal toxicity, and acute phase reactions are commonly seen after the first infusion. The long-term possibilities such as growth retardation (in animal models), iatrogenic osteopenosis, esophageal cancer, delayed fracture healing, and osteoradionecrosis of the jaw are particularly disconcerting. Dental side effects include delayed tooth eruption (in rats), enamel defects and areas of ankyloses between the alveolar bone and the developing hard dental tissues. Long-term studies are required to conclusively determine the safety of these drugs.

FD and malignancy

The evolution of FD cannot be gauged with any biomarkers. This is particularly disconcerting in pediatric patients due to active growth. The rapidly progressing nature and subsequent cystic degeneration of the disease may also be confused with malignant transformation.

The first case of malignancy arising from FD was described by Coley and Stewart in 1945. Malignancies are rare in FD but may be observed both in monostotic and polyostotic forms. In a large series, the frequency of malignant transformation varies between 0.5% and 4%, although this incidence may have been overestimated because many of the patients had received radiotherapy. The tumor is usually an osteosarcoma rather than fibrosarcoma or chondrosarcoma. The majority of the patients who presented with malignancy were beyond the fourth decade (80%) and the mean age at the time of occurrence being 46.5 years. An accurate clinical history is vital to early recognition of a malignancy developing in FD. The patients almost invariably have rapidly developing symptoms such as pain and swelling. The most constant radiographic feature is the permeation of the lesion through the bony cortex and subsequent involvement of the surrounding soft tissue. Surgically treated cases with positive or uncertain margins should be subjected to post-operative radiotherapy or chemotherapy. reported a poor prognosis with a 53.6% mortality rate. Studies have also suggested that surgical intervention in the form of internal fixation or bone grafting provides slots in which mesenchymal cells develop tumors. However, there was not enough evidence support this theory.

Conclusion

Craniofacial FD is a progressive disease which necessitates the removal of all dysplastic tissue failing, which severe complications could occur. This, however, is easier said than done as the resulting morbidity could eclipse the surgical achievement. Controlling the disease by minimal surgical intervention during the symptomatic phases seems like the prudent approach for pediatric cases. In our case, the bony enlargement on the lateral wall had enlarged to an extent that caused proptosis, epiphora, and pain. We believed that vision was in imminent danger and prophylactically decompressed the canal while contouring the lateral wall. Well-designed trials are required to study genetic manipulation which could possibly revolutionize the treatment of this disease.

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