

SUCCESSFUL MANAGEMENT OF TRISMUS IN A RARE CASE OF FIBROUS DYSPLASIA OF THE CORONOID PROCESS

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ABSTRACT

Fibrous dysplasia (FD) is a slowly progressive benign bone condition in which normal bone is replaced by an abnormal fibro-osseous tissue. FD has various manifestations and can present with significant cosmetic and functional disturbances, particularly in the craniofacial skeleton. We present a craniofacial fibrous dysplasia (CFD) case in a 6 year old child who presented with severely reduced mouth opening and some aesthetic concerns. The patient underwent left side coronoidectomy and contouring of the involved facial skeleton through bicoronal approach. There was significant improvement in mouth opening. The patient was very satisfied with the outcome. This case report highlights the importance of individualized treatment plan addressing the patient's concerns.

Key words: Fibrous dysplasia–Craniofacial fibrous dysplasia–McCune-Albright syndrome

1 INTRODUCTION

Fibrous dysplasia is a slowly progressive, expansile, benign bony disorder in which normal bone is replaced by abnormal fibro-osseous tissue [1]. The etiology of this abnormal growth process is a mutation in the gene encoding the subunit of a stimulatory G protein (G_{α}) located on chromosome 20. This mutation leads to a substitution of the cysteine or histidine amino acids of the genomic DNA by arginine in the osteoblastic cells [2]. Consequently, the bone is transformed into cellular fibrous tissue containing irregular trabeculae. It represents 2.5% of all bone tumors and 7% of benign bone tumors [2].

Von Reclinghausen in 1891, initially described it as “osteitis fibrosa generalisata”, in a patient with skeletal deformities due to fibrotic bone changes. Later in 1938, Lichtenstein introduced the term “Fibrous dysplasia” for this

disorder. Fibrous dysplasia (FD) is described as three major types: monostotic, involving a single bone; polyostotic, having multiple lesions involving multiple bones; and craniofacial, confined to bones of the craniofacial skeleton. McCune Albright syndrome, a variant of polyostotic fibrous dysplasia is associated with cafe-au-lait pigmentation, and endocrinopathies including hyperthyroidism and sexual dysfunction [3]. The prevalence of FD for monostotic type is 74%, for polyostotic type is 13% and for craniofacial type is 13% [4]. Most cases of craniofacial FD cannot be categorized as monostotic because of the involvement of multiple adjacent craniofacial skeleton and also, they are not truly polyostotic because bones outside craniofacial complex are also not involved [5]. The sites of skeletal involvement are manifested early in patients with fibrous dysplasia. Around 90 % of craniofacial lesions are established before 5 years of age, and 75 % of all sites of FD are expressed by 15 years of age [6].

In the face, the common complaints are asymmetry and swelling. Depending on the site and bone involvement, other

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symptoms include nasal obstruction, when the paranasal sinuses are involved; visual disturbances if the orbit is involved; hearing loss when a temporal bone lesion is present [3]. The growth is progressive and may impinge on other structures leading to functional impairment.

Here, we report a case of craniofacial dysplasia in a 6 year old male patient, who presented with reduced mouth opening and facial asymmetry and was successfully managed with left coronoidectomy and contouring of the involved skeletal elements.

2 CASE REPORT

A 6 year old male child presented to the plastic surgery department with reduced mouth opening and multiple swellings of the frontal bone on the left side for the past 1 year. Because of the limited mouth opening, patient was unable to take solid food since 1 year and was on liquid diet. He had lost significant weight over the last 6 months. He had been operated twice earlier at the age of 2.5 years and 4 years for the frontal bone swellings. Extraoral examination revealed facial asymmetry with bony hard swelling of the left maxilla. Obvious swellings on the frontal bone on left side and glabella created an unaesthetic appearance (Figure 1). He had mild hearing loss on the left side. Intraoral examination revealed poor oral hygiene with multiple carious teeth and mouth opening of only 8 mm. There was no swelling present elsewhere in the body.

The computed tomography (CT) showed radiodense mass with ground glass appearance involving left maxilla, left coronoid process, left fronto-parietal region, left temporal region suggestive of fibrous dysplasia (Figure 2,3) Contact between the enlarged left coronoid process and the zygomatic arch, limited the mouth opening. Based on clinical history and radiographic assessment, a provisional diagnosis of craniofacial fibrous dysplasia was made. Surgical plan was discussed with neurosurgery team and included left coronoidectomy with removal of zygomatic arch to improve mouth opening and contouring the frontal bone and glabella prominences.

SURGICAL TECHNIQUE

We started with bicoronal incision aimed at surgical contouring and debulking of abnormal bone in the frontal region. The incision was given and scalp flap was reflected to expose bone from coronal sutures to bilateral orbital margins. Debulking was done using rotatory instruments. Irregular surface protuberances were smoothed. The contour and aesthetics of the region was evaluated by putting back the scalp flap and reviewing the appearance time to time. The symmetry of the contour in frontal region was achieved satisfactorily.

The bicoronal incision was extended in the preauricular region upto the lobule of ear to approach the left coronoid process (Figure 4). In the temporal region, this incision was upto the superficial layer of the temporal fascia. At the root of the zygomatic arch, the superficial layer of the temporalis fascia was incised antero-superiorly at a 45° angle.

The periosteum was incised to expose the zygomatic arch. A transzygomatic approach was planned to access the enlarged coronoid. Around 1.5 cm of the zygomatic arch was excised. The coronoid process was exposed and the temporalis muscle attachment was stripped. Osteotomy was made extending from the depth of sigmoid notch to the anterior border of the ramus. Mouth opening of 30 mm was achieved. Bony margins were smoothed. A suction drain was placed and flap was closed in layers. 1 cm incision was given over the medial canthal swelling and underlying bone was contoured. Medial canthopexy was done using 4-0 prolene. The bone chips were sent for histopathological examination.

Postoperative course- The postoperative course was uneventful. Vigorous physiotherapy was started from third postoperative day with wooden spatula to maintain the mobility of the joint and prevent relapse. At 2 weeks postop, the mouth opening was 27 mm (Figure 5). The patient was trained and encouraged to continue mouth opening exercises 4-5 times a day for the next 6 months. He was referred to the dentist for the oral rehabilitation.

Histopathologic examination showed trabeculae of lamellar bone with intervening fibrous tissue confirming FD.

After 2 months postoperatively, he maintains excellent mouth opening and has gained some weight. The family is very satisfied with the outcome



Figure 1. Preoperative clinical photograph of the patient revealing facial asymmetry

3 DISCUSSION

Fibrous dysplasia is a benign condition in which normal bone is replaced by abnormal fibrous tissue and haphazardly distributed woven bone [7]. FD exists along a broad clinical spectrum, ranging from an isolated, trivial lesion to severe disease involving nearly the entire skeleton. Clinically

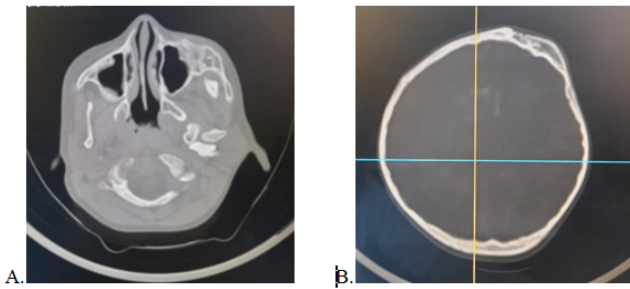


Figure 2. Axial computed tomography slice with bone window revealing involvement of left maxilla (A) and left side frontal bone (B)

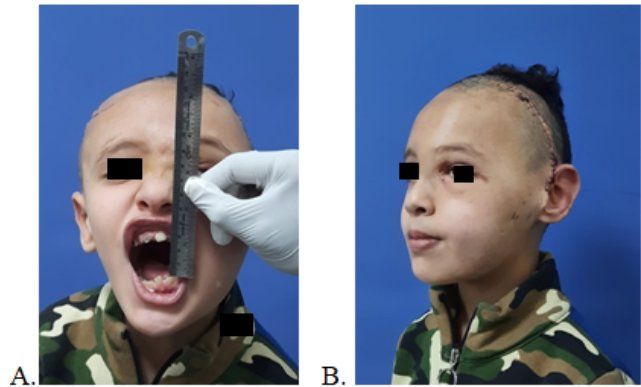


Figure 5. 1 week postoperative photograph showing A) Increased mouth opening B) Satisfactory aesthetics

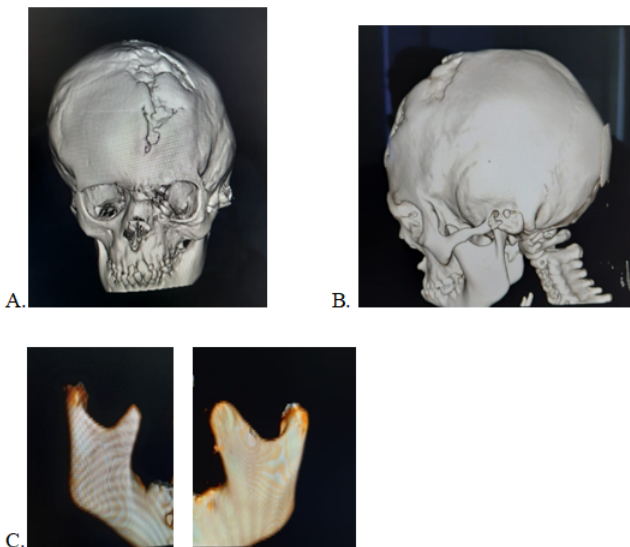


Figure 3. Three dimensional CT reconstruction shows A) Involvement of the left side frontal bone, left maxilla, glabella B) Enlarged left coronoid process C) Grossly enlarged left coronoid process as compared to right coronoid process

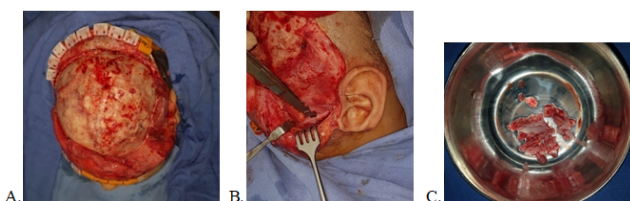


Figure 4. Intraoperative photograph showing A) Coronal approach for surgical contouring B) Preauricular extension on the left side to expose enlarged coronoid process C) Surgically removed fibrous dysplastic bone pieces

significant bony lesions are usually apparent by age 5 years [8]. Craniofacial FD lesions are some of the earliest to be detected and involves the maxilla almost twice as often as the mandible and is usually unilateral in nature [5].

FD in the craniofacial region can cause significant expansion of bones, with varying degrees of facial asymmetry and disfigurement. The signs and symptoms of FD are variable and are dependent on the location of the lesion and its compressive effect on neighboring structures as the tumor slowly progresses [9]. Although the lesion is usually asymptomatic, but when it encroaches on canal or foramina, it causes considerable pain and discomfort [10]. In the craniofacial region, it usually begins as an asymptomatic painless swellings leading to facial asymmetry and depending on the site and bone involvement, other symptoms include nasal obstruction when paranasal sinuses are involved, visual disturbances if the orbit is involved, whereas temporal lesions may produce hearing loss [3].

The frequency of malignant transformation of FD is 0.4-1% and the interval from development to malignancy is usually years to decades. The most common malignancies are osteosarcoma and fibrosarcoma. Features suggestive of malignant degeneration include rapid increase in the size of the lesion, increasing pain and rapidly enlarging soft tissue mass [2].

Computer tomography (CT) is a superior diagnostic tool and invaluable in pre-operative planning. FD has characteristic appearances on CT and consists of 3 varieties: ground glass pattern (56 %), homogeneously dense pattern (23 %) and cystic variety (21 %) [2]. Introduction of 3-dimensional CT improves the localization of the pathology and assists in accurate surgical planning. It can also serve as a precise baseline study for future follow up comparisons.

Magnetic resonance imaging (MRI) may also help in evaluating cranial nerve involvement and soft tissue structures adjacent to the lesion [3]. On MRI, fibrous dysplasia exhibits homogenous, moderately low signal intensity on T1 weighted images. On T2 weighted images, the tissue usually exhibits very high signal intensity [11].

Bone scintigraphy, usually ^{99m}Tc-MDP is exquisitely sensitive for detecting FD lesions and is usually recommended

to rule out the polyostotic variant of FD. It is a useful imaging modality for detecting the distribution of lesions, and has been useful for discriminating between monostotic and polyostotic disease, ruling out metastasis of secondary sarcoma, and recurrence after therapy [12].

Histopathology is confirmatory in diagnosing FD and shows cellular connective tissue stroma comprising of woven bone arranged in various shapes of trabeculae. The definitive diagnosis of FD is made by correlation of clinical, radiological and histopathological findings.

The mere presence of CFD is not an indication for treatment as many small solitary lesions will remain asymptomatic for long periods [2]. Functional issues should take precedence over esthetic issues in the management of FD. The choice of surgical option depends on several factors: site of involvement, rate of growth, functional disruption, aesthetic disturbance, age and general health of the patient, patient's preference, surgeon's experience and availability of a multi-disciplinary team including neurosurgeon, craniofacial surgeon, ophthalmologist and pediatric dentist [2].

Surgical contouring or resection may be warranted prior to skeletal maturity if there are symptoms, rapid change in the lesion or function is affected, however, patient must be aware of the risk of regrowth. An individualized strategy should be adapted to the patient's functional and aesthetic complaints, aiming at aesthetic correction or functional improvement. In the present case, patient had limited mouth opening because of the impingement of elongated coronoid process to the posterior aspect of zygomatic process at the mouth opening and had multiple swellings of the frontal bone and glabella and hence, coronoidectomy was done to improve the mouth opening and surgical contouring of the frontal, glabella protuberances was done for aesthetic correction.

Enlarged coronoid process leads to progressive limitation in mouth opening by impinging upon the medial surface of the zygomatic arch. Progressive impairment of mouth opening interferes with eating, speaking and maintain oral hygiene. The definitive treatment for enlarged coronoid process is coronoidectomy. If the mass is large and in close proximity to the zygomatic arch, an extraoral approach allows better access and visualization [13]. Postoperative physical therapy is very important for obtaining a good result after coronoidectomy. Active physiotherapy should commence immediately after surgery and patient should be instructed to perform the exercise several times a day for at least 6 months for satisfactory results.

Medical treatment with bisphosphonates such as alendronate or pamidronate has been used to reduce pain and inflammatory symptoms, reduce bone destruction, increase the osseous density and reduce the growth of the lesions [3]. Serum alkaline phosphatase, a bone turnover marker, is consistently reduced in patients treated with bisphosphonate to monitor the response to medical treatment.

Radiotherapy is contraindicated in FD because of the possibility of subsequent development of radiation-induced sarcomas [1]. Radiation therapy elevates malignancy risk by 400-fold [14]. Regular follow-up is of fundamental importance in order to detect relapses or a possible, malignant

change at an early stage.

4 CONCLUSION

Fibrous dysplasia is considered a bony pathology which may present with functional and aesthetic impairment. Surgical treatment remains the cornerstone of treatment for CFD. Conservative debulking and recontouring procedures aiming at functional improvement and aesthetic correction may be warranted prior to skeletal maturity if there are symptoms or rapid change in the lesion. After coronoidectomy, long term physiotherapy is important and should be initiated in early postoperative phase. Craniofacial surgeries should be planned utilizing a team approach addressing the patient's specific concern and the patient should remain under regular follow up.

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