Fibrous Dysplasia in the Craniofacial Region: a retrospective review of cases treated in a Tertiary Hospital, North- west Nigeria

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Abstract

Background: Fibrous dysplasia (FD) is a rare bony disorder in which normal bone is replaced by abnormal fibro-osseous tissue. It often involves the long bones, craniofacial bones, ribs, and pelvis. It often occurs in the first and second decades of life. We, therefore retrospectively review cases of fibrous dysplasia of the craniofacial region treated in our Centre.

Objective: To present the clinical features, radiological appearances and treatments of craniofacial fibrous dysplasia.

Methodology: The study was a retrospective review of case files of patients that were treated in our Centre for fibrous dysplasia of the craniofacial region from January, 2009 to October, 2021. Records of patients were obtained from clinic register and operation register. The case folders of the patients were retrieved and analyzed for age, sex, site, clinical features, radiological findings and treatment received.

Results: Thirty-three cases were included in the study. Majority of the cases occurred in the age group 11 – 20 years (n=22, 66.7%), the least affected was age group 51 - 60 (n=1, 3.0%). Females were more affected (n=20, 60.6%) than males (n=13, (39.4%) at the ration of 1.5:1. The maxillae was most affected (n=21, 63.7%), then mandible (n=11, 33.3%), the cranial bones were least affected (n=1, 3.7%). Surgical excision was the modality of treatment for all our cases.

Conclusion: Fibrous dysplasia causes facial disfigurement and surgical excision is the ideal treatment with excellent outcome. CT scan, radiographic examination of the lesion before surgery and histopathology of excised tissue are crucial to its diagnosis.

Keywords: Fibrous dysplasia, mandible, maxillae, fibro-osseous, cranial bones.

Introduction

Fibrous dysplasia (FD) is a rare, non-malignant condition in which normal bone and marrow are replaced by fibrous tissue and randomly distributed woven bone.[1] The tumour is usually accompanied with pain, bony deformity, and pathologic fractures.[1,2] Lichtenstein in 1938 first named the lesion fibrous dysplasia (FD).[1] FD presents clinically in three forms namely: monostotic (single bone affected) , polyostotic (multiple bones affected), and polyostotic with endocrinopathies, which can be associated with hyperpigmentation and endocrinological disorders and is called McCune-Albright syndrome.[3] This tumor is mainly bony disorder and common sites of skeletal involvement are: the long bones, craniofacial bones, ribs and pelvis. Monostotic FD (MFD) has relatively high frequency of occurrence in the jaws compared to the other types of FD.[4]

However, FD is caused by somatic activating mutations of the gene GNAS in a subunit of the stimulatory G protein, located at 20q13.213.3.[5,6] The diagnosis of FD is based on clinical, radiological, and histopathological examination. A radiological feature shows a ground glass appearance. Many researchers[5-7] reported that FD is commoner in teenagers, and it usually becomes static after adulthood.[5] It commonly affects females more than males.[6] Jaw lesions cause displacement of teeth, malocclusion, loss of lamina dura, narrowing of the periodontal ligament space, and rarely root resorption.[4] Nasal obstruction may occur if paranasal sinuses are affected. Lesions extending to the orbit may cause visual impairment and temporal bone lesions may cause hearing loss.[4,5] Facial pain, headaches, or facial numbness may develop more in craniofacial FD.[5]

Moreover, FD involves the maxilla almost two times more often than the mandible.[4-6] It frequently appears in the posterior region of the jaw bone and is usually unilateral.[5] There are different treatment approaches to FD which include observation, medical treatment, and surgical treatment. This study reviewed cases of craniofacial FD treated in Barau Dikko Teaching Hospital, Kaduna, North-west, Nigeria.

Methodology

This was a retrospective study of cases of craniofacial fibrous dysplasia in the skull and jaws that were treated at the Maxillofacial Clinic, Barau Dikko Teaching Hospital, Kaduna, Nigeria, from January, 2009 to October, 2021. The sample frame was thirty-five. The sample size was thirty-three. Two patients whose records were inadequate were excluded from the study. Records of patients were obtained from clinic register and operation

register. The case folders of the patients were retrieved and analyzed for age, sex, site, clinical features, radiological findings and treatment received. Data were sorted, organized and entered into SPPversion 20(IBM^0 SPSS^0 statistics Armonk New York, United States) for analysis. Frequency statistics and cross tabulations were done and chi-squared test was used to test for significance between variables at the critical $p<0.05$.

Clinical presentation of FD: Facial asymmetry was a common presentation, both labial and lingual bone expansion in the maxillae and mandible were common (Figure 1). Malocclusion was observed in few cases. The overlying mucosa had normal color and appearance. One case of polyostotic FD with cranial bones involvement (temple and occiput), also had multiple skin pigmentations but no abnormal neurological findings.

Radiological features: Radiographic examination revealed a lesion with both radiopaque and radiolucent features showing a ground-glass appearance, there was buccal and lingual bone expansion. Panoramic radiography and plain radiographs of the jaws were all useful in the diagnosis of FD. Computerized Tomography scan was a useful imaging technique for CFD. CT scan of CFD imaging showed expansive mass with ground-glass opacity involving the mandible, maxilla, temporal bones and occiput.

Histological features: Macroscopic features showed a grayish solid mass, Histopathology revealed that the tumor was composed of a solid proliferation of spindle-shaped cells associated with islands of osteoid and bone trabeculae. The trabeculae of woven bones had irregular size, form, and distribution.

Results

TABLE 1: AGE DISTRIBUTION

AGE/ YEARS	NO OF CASES	PERCENTAGE
11-20	22	66.7
21-30	5	15.1
31-40	3	9.1
41-50	2	6.1
51-60	1	3.0
TOTAL	33	100

TABLE 2: SITE AND SEX DISTRIBUTION

SITE	SEX		TOTAL NO OF CASES	PERCENTAGE BY SITE
	M	F		
Maxillae	9	12	21	63.7
Mandible	4	7	11	33.3
Cranial bones	-	1	1	3.0
Total	13	20	33	100

Thirty-three cases cases were included in the study. Majority of the cases occurred in the age group 11 – 20 years (n=22, 66.7%), the least affected was age group 51 - 60 (n=1, 3.0%) (Table 1).

Females were more affected (n=20, 60.6%) than males (n=13, (39.4%) at the ratio of 1.5:1, The maxillae was most affected (n=21, 63.7%), then mandible (n=11, 33.3%), the cranial bones were least affected (n=1, 3.7%) (Table 2). Surgical excision was the modality of treatment for all our cases.

Figure 1a: A 16 -years old boy with maxillary tumour

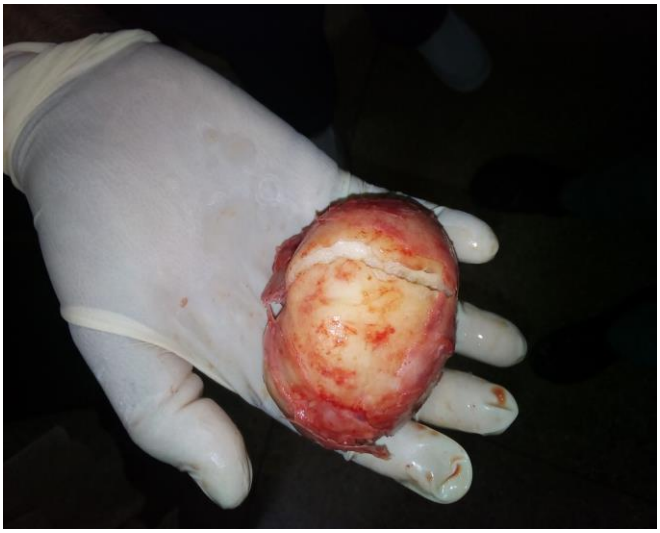

Figure 1b. The excised tumour

Figure 2a. A 12-years old girl with tumour of the maxillae

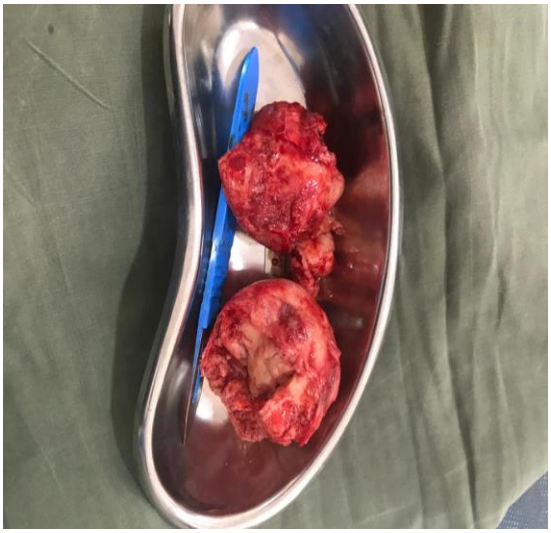

Figure 2b. The excised Tumour

Discussion

Fibrous dysplasia (FD) is a benign fibro-osseous bone dysplasia that can involve single (monostotic) or multiple (polyostotic) bones.[7] Monostotic form is more frequent in the jaws.[9] It is termed as craniofacial fibrous dysplasia, when it involves, though rarely, adjacent craniofacial bones. Majority of our cases were monostotic (96.9%) involving either the mandible or the maxillae. One case of polyostotic FD (3.1%) was reported in our study. This further supported other researchers that reported rarity of polyostotic type.[5,6]

The maxilla is more commonly involved than the mandible in monostotic jaw bone lesions.[8,9] This study reported that the maxillae was more affected than the mandible in the ratio of 1.9 to 1.0 (Table 2). Our cases that involved the mandible were located more in the anterior region than posterior. The maxillary lesions were more in the posterior region (Figure 2). Most of our cases had bucco-lingual expansion either in the maxillae or the mandible. The lesion presented with varied sizes which ranged from few millimeters to massive tumor. The most common symptoms of our cases were: facial deformity with asymmetry, nasal congestion, malocclusion, dental anarchy, pains and few with pus discharge from the lesion (Figure 1). FD can occur at any age, but more common in children and young adults.[9,10] However, our study showed that children in their second decade of life were most affected (66.7%), but elderly were also affected but very low incidence (Table 1). Several authors[4-7] had reported that FD had female predilection, our study supported this claim with the report of female to male ratio of 1.9 to 1.0. However, in some other studies, no gender predilection was found.[11,12]

Moreover, some researchers [11,12] had claimed that FD progress slowly and ceases after puberty or bone maturation, whereas others[7,9,10] reported, that FD continued to progress into old age. This study could conclude that FD progresses to old age as one of our female cases presented at age of 56-years with a maxillary lesion.

However, the radiographic appearances of FD of the jaws in this study ranged from radiolucent to radio-opaque, mature tumor showed waves of radio-opacities appearing in ground glass pattern. Several authors[7,8] had reported three different radiographical patterns which were: cystic (radiolucent or lytic), sclerotic, and mixed (radiolucent/radiopaque). Our study also agreed with this findings, early lesions presented as radiolucent lesion, while those lesions that were age 2 to 4-years duration presented as mixed radiolucent and radio-opaque, long duration of 12-years were hyperdense radio-opaque (sclerotic).

The differential diagnosis of FD includes: osteoma, ameloblastoma, cementoma, cementifying fibroma, benign odontogenic tumour, simple bone cyst, non-ossifying fibroma, osteofibrous dysplasia, low grade intramedullary osteosarcoma, and Paget's disease. The diagnosis of FD is confirmed with histology, but radiographic findings could be of great assistance. Treatment protocols for FD as suggested by an author[14] include: observation, medical treatment, and surgery. Clinical observation is suggested for FD lesions that have no risk of pathologic fracture or facial deformity.[14] Medical treatment with bisphosphonates may have benefits including improvement of function, pain relief, and lower fracture risk for appropriately selected FD patients.[14,15] All our cases had radical excision of the tumor to prevent recurrence. Tumor recurrence was reported in two of our cases. The first excised tumor gave histolology of FD, but after second surgery the histology was ameloblastoma. This greatly proved that histological diagnosis was not absolutely reliable. There were associated morbidities following surgery, such as oro-antral fistula, loss of dentoalveolar segment and phonation defect. Prosthesis was fabricated for the patients to improve oral function defects. In conclusion FD could be described as non-life threatening tumour as all our cases had complete recovery from the lesion.

Limitations: Patients were not keeping to follow up appointment which made long term evaluation of our treatment modalities impossible. All our cases were treated with surgery whereas there are other modalities of treatment such as observation, medication with bisphosphonates and surgical reduction of the tumor.

Consent: Patients gave their consent for their images and other clinical information to be reported in the journal.

Conflict of interest: None

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