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Case Report

Craniofacial polyostotic fibrous dysplasia: A rare case

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ABSTRACT

Craniofacial fibrous dysplasia (FD) is one of three types of FD which can affect the craniofacial complex. It is the proliferation of cellular fibrous connective tissue intermixed with irregular bony trabecules. It is a developmental tumor-like condition that is characterized by replacement of normal bone. The purpose of this report is to present a rare case of craniofacial polyostotic FD that led to vision loss in the same affected side.

Key Words: Craniofacial, non syndromic, polyostotic fibrous dysplasia

INTRODUCTION

Craniofacial fibrous dysplasia (FD) is a bone development anomaly characterized by hamartoma proliferation of fibrous tissue within the medullary bone, with secondary bony metaplasia, producing immature, newly formed and weakly calcified bone without maturation of osteoblast which appears on radiographs as a radiolucent, mixed, or radiopaque lesion.[1]

FD is a benign and slow-growing lesion. The most common presenting symptom is painless enlargement. In most cases, a specific diagnosis cannot be made on a histological basis alone, nor can it be made without appropriate radiographs of high quality.[2-4]

Involvement of the face and skull by FD could be mentioned, which is known as "leontiasisossea".[5] It tends to expand during childhood and stabilize after puberty.[6,7] There is no sex inclination. It may be seen as monostatic or polyostotic type. Sometimes, it is associated with abnormal endocrine hyper function and pigmented skin lesions, which is known as "Maccune-Albright" syndrome.[8]

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Cranial bones are common places for involvement with 10 to 27% in monostatic and 50% in polyostotic cases.[9,10] Temporal bone involvement is less common.[11,12] Depending on the type and location of FD, the signs and symptoms vary and include facial deformity and asymmetry, vision changes, hearing impairment, nasal congestion and/or obstruction, pain, paresthesia, and malocclusion.[13]

In this paper, a case of craniofacial polyostotic FD in a 10-year-old boy is described. Prominent properties of this case could be mentioned as: Extensive involvement of left hemicranium, progressive course of disease, especially proptosis in left eye and frontal bossing due to the large growth of diseased bone and occurrence in temporal bone.

CASE REPORT

A 10-year-old boy was referred to our department with complaint of painless swelling in the left side of the skull and face since he was 3 years old. Swelling was small at first and gradually reached to present extension [Figure 1].

In extra oral examination, diffused expansion was seen on left side of his face. The lesion displaced his left eye and led to exophthalmos. Frontal bossing was seen on the left side. Surface temperature of skin was higher in left side compared to right side. A non-tender mass was palpated on his left temporal bone. Consistency was bony hard [Figures 2-4].

In intra oral examination, a bony expansion was found on buccal surface of maxilla from distal of lateral incisor to distal of first molar. Horizontal dimension was approximately 4 cm. Palatal aspect wasn't expanded. Overlying mucosa was intact and non-tender [Figure 5].

There were no signs of precocious puberty. Cutaneous pigmentation or any endocrinological disorder was not detected in the physical and laboratory examinations. His vision in left eye was lost 2 years before in a firecracker incident.

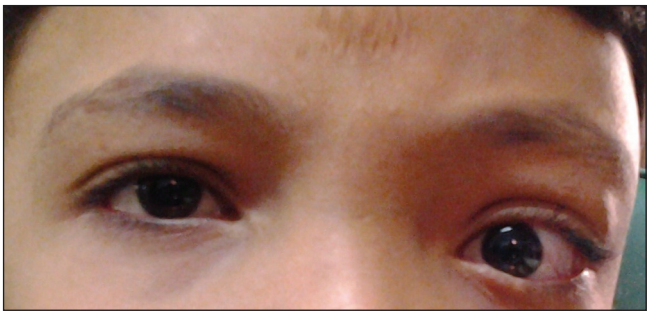

Figure 1: Clinical view of patient

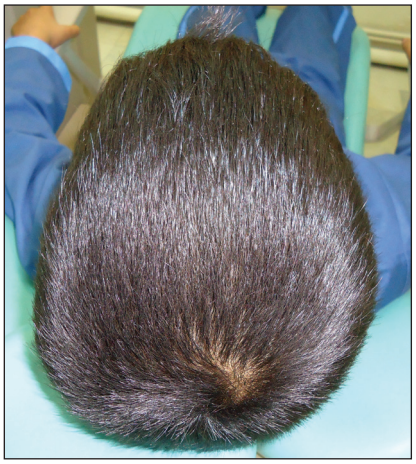

Figure 3: Occipital involvement of cranial

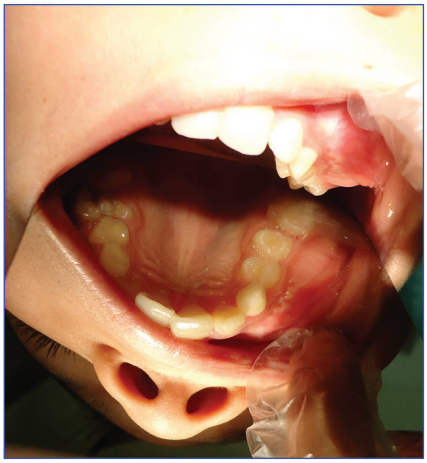

Figure 5: Intraoral view of lesion was extended from distal of lateral to distal of first molar

Radiological findings

In an initial panoramic image, the vertical height of the body of the mandible in this area seemed to be larger in comparison with the right side. There was a generalized ground glass appearance in left maxilla and mandible quadrant [Figure 6].

Cranial and facial bones were evaluated with computerized tomography (CT) scan in axial and coronal

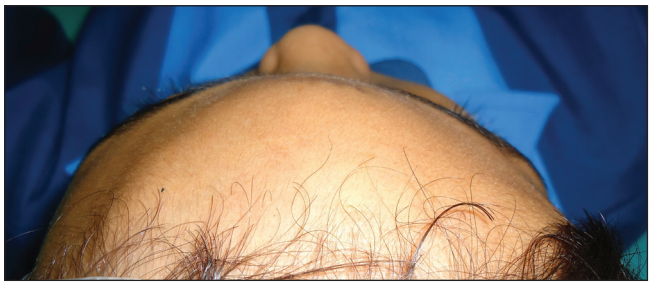

Figure 2: Frontal bossing

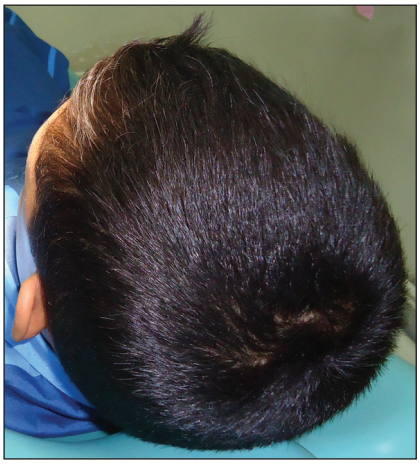

Figure 4: Swelling of left side

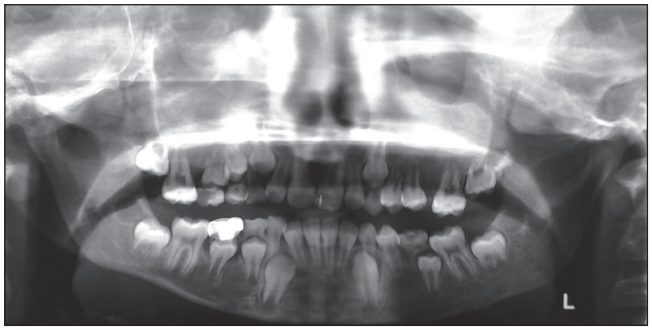

Figure 6: Panoramic view shows widening of the body in left side

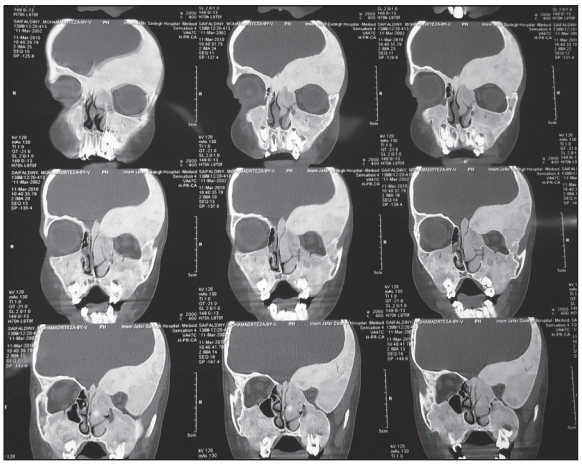

Figure 7: CT scan in coronal plans. Cranial CT scan showed extensive involvement of most cranial bones in left side

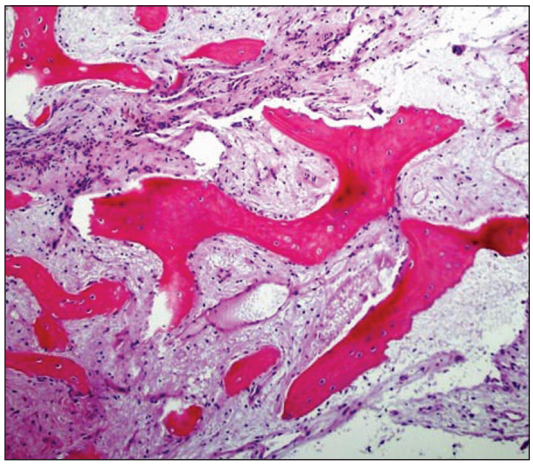

Figure 8: Histopathologic view of lesion in incisional biopsy

plans. Cranial CT scan showed extensive involvement of most cranial bones in left side [Figure 7].

Laboratory findings

Because of high temperature of head and face in affected side compared to other side, a list of laboratory tests was requested. Ca level was 10.5, ALP was 1424, and phosphorus was 4.2. These findings affirmed increase bone metabolic activity.

Pathological findings

Poorly oriented trabeculae of bone were separated by cellular fibrous connective tissue. The bone appeared woven rather than lamellar [Figure 8]. Chinese alphabet pattern was seen that confirmed FD [Figure 8].

DISCUSSION

FD is one of the several fibro-osseous lesions that affect craniofacial region. This lesion-like cherubism and focal cement osseous dysplasia can occur anywhere in the skull. But occurrence in maxillofacial region is more usual.[14] Ground glass and orange peel appearance are the main radiographic pattern of FD.[15]

This lesion have wide spectrum of radiographic patterns from completely radiolucent, mixed to completely radiopaque.[16]

Hence, other lesions like cemento ossifying fibroma (COF) can imitate this radiographic pattern.[17] It is critical for the clinician to consider it among differential diagnosis.[18]

In most cases, the radiographic characteristics of polyostotic FD and clinical information are sufficient to render definite diagnosis without biopsy. Usually, diagnosis of polyostotic type opposed to monostatic type is based on clinical behavior.[15]

Because cherubism is a bilateral lesion growing without any neurological damage or deformity in the face, it wasn't a reasonable diagnosis.[17] Among the other diagnosis was focal cemento osseous dysplasia (FCOD). Although FCOD is a maturable lesion, it usually affects mandible in single form and affects more quadrants in both sides. But, in this case, the lesion affected left side of both the jaws. Also, FCOD usually appears in females in fourth decade of life and not in children.[18]

COF is a fibro osseous lesion which can show a ground glass pattern, but it usually occurs in females and it doesn't have skull involvement. For definite diagnosis, an incisional biopsy was achieved from buccal surface of expanding lesion on left maxillary quadrant.

CONCLUSION

FD may manifest as monostotic or polyostotic forms. In polyostotic form, it is important to monitor growth of lesion for possible first step surgery to prevent neurological defect or face deformity.

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