Fibrous Dysplasia of Maxillary Bone

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Abstract

Fibrous dysplasia is an idiopathic developmental bone disease of benign origin that has been linked to a localized developmental arrest; all of the components of normal bone are present but they do not differentiate into their mature structures. It causes skeletal deformity & pathological fracture of bone. The lesion appear in three distinctive but sometimes overlapping clinical pattern 1.single bone involvement(monostotic);2.multiple bone involvement (polyostotic);and 3. polystotyic disease, associated with café-au-lait skin pigmentation and endocrine abnormalities, especially precocious puberty. The study was done in CBMCB hospital. The patient came to us with a swelling in the left side of check and a mass in left nasal cavity. Biopsy was taken from nasal mass which revealed fibrous dysplasia. Later on, surgical intervention was done by lateral rhinotomy approach. Post operative period was uneven full and patient was discharged on 7th POD. After four weeks interval, follow up revealed no deformity in check .No nasal obstruction was found and scar mark was also reasonable.

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Key words: Fibrous dysplasia, Maxilla, Monostotic form.

Introduction

Fibrous dysplasia is not an uncommon tumour like condition. It causes expansion and replacement of medullary bone by disorganized fibroosseous tissue[1]. It comprises 2.5% of all osseous & 7% of all benign bone tumours[2]. In general they affect 1 in 4000 to 10000 individuals[3]. Male are affected as commonly as females[4] . It is usually present before the third decade of life[5]. usually an early adolescence.

Monostotic fibrous dysplasia accounts for 70% of all cases. Often stop enlarging at the time of growth plate closure. Most ffrequently found in metaphysodiaphyseal region of long bones i.e. rib (28%),followed by femur (23%),tibia, craniofacial bone (10-25%)[6].Malignant transformation has been reported in 0.5% of patients[7].A more aggressive form of the disease, Mc Cune-Albright syndrome, is associated with a traid of café-au-lait spots, precocious puberty and fibrous dysplasia.[8,9].In 10% of monostotic fibrous dysplasia and 50%-100% of polyostotic fibrous dysplasia, there is involvement of the facial and cranial bones [1,4].

Case report

A 12 years old girl got admitted in Community Based medical college hospital with the complaints of left sided nasal obstruction for 8 months and discharge from the right side of the nasal cavity. She also noticed swelling over the left side of the nose and check for 3 months and which was gradually increasing in size. Local examination reveals a non tender swelling over the dorsum of the nose and left maxillary region (Fig. 1).

Anterior rhinoscopy showed a reddish mass in left nasal cavity which was firm in consistency , sensitive to touch, nontender and seemed to be arises from lateral wall of the nose. X ray PNS shows opacity in left maxillary sinus and septum was deviated to right side. CT scan of PNS with contrast revealed-expansile soft tissue density with in adjacent it maxillary, frontal, ethmoidal and sphenoidal sinuses.

Biopsy taken from the nasal mass revealed features of fibrous dysplasia. The operation was done on 14/09/2011. Surgical procedure- Orotracheal intubation was done , followed by after proper toileting of the operative area patient was drapped properly. Then lateral rhinotomy incision was given in left side. After proper exposure the mass was removed from nasal cavity as well as from left maxillary sinus (Fig. 2).

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It was firm to hard in consistency . After cleaning the maxillary sinus it was found that the ethmoidal and sphenoidal region was full of fibrous variety mass which also excised out and haemostasis ensured. Then ribon gauge soaked with antibiotic pack was given .Wound was repaired in two layers, inner layer with 3.0 vicryl then skin with 4.0 prolene. Pre & post operatively inj ceftriaxone bid was given for a period of 5 days. Post operative period was unevenful. Nasal pack was removed after 48 hours. On 5th POD stitches was removed alternatively and on 6th POD all stitches removed. Patient was discharged on 7th POD (Fig. 3).

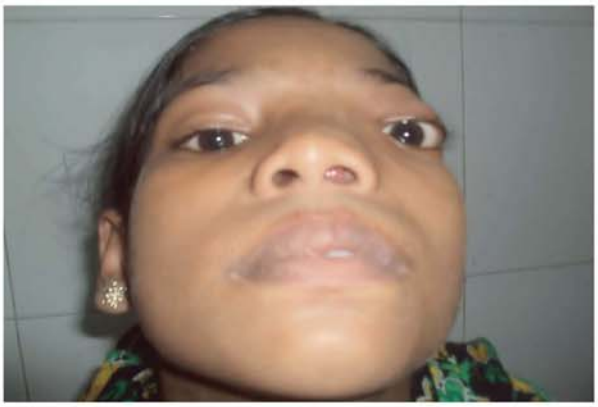

Fig.: 01

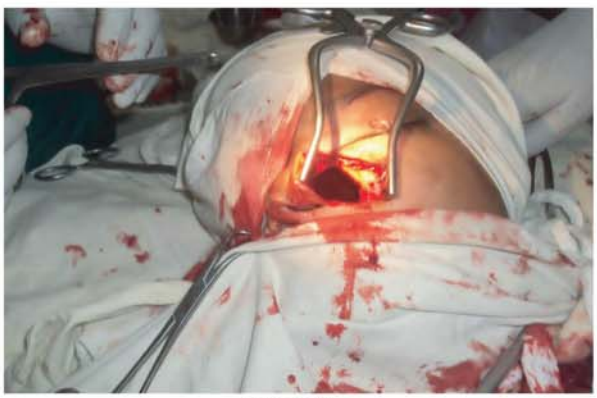

Fig.: 02

Fig.: 03

Specimen sent for histopathological examination which showed densely packed, irregular shaped trabecule of woven bone surrounded by a moderately cellular fibroblastic cells proliferation.

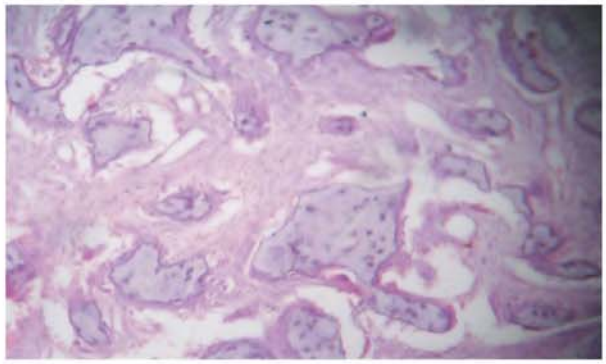

Fig.: 04 showing fibrous dysplasia within the bone tissue

Discussion

Fibrous dysplasia results from post zygotic mutation in GNAS1(Guanine Nucleotide Binding Protein ,α -stimulating activity polypeptide-1) gene[4,6].Clinical severity depends on time when the mutation of GNAS-1 occurs. If mutation occur during post natal life the progeny of that mutated cells are essentially confined to one site resulting in fibrous dysplasia affecting a single bone.

The disease is progressive but with time will often burn out and stabilize. Fibrous dysplasia may stabilize after puberty. [10].The disease may progress into adulthood[11] And observing the patient for several years before cosmetic surgery is underlies is prudent [12,13].Progression in later life may occur [11,14].The type of surgery performed varies from shaving and contouring of the bone to radical surgery. Complete resection may be required in patient with Albright syndrome , science there is a high incidence of progressive growth and recurrence[13].For large cosmetic deformities, recontouring and repositioning of bone may be required. Orbital complication like loss of vision is one of the most severe but relatively uncommon complication[14].Surgical intervention is indicated for visual impairment caused by fibrosis dysplasia but prophylactic decompression is controversial [15,16].Medical therapy with bisphosphonates may stabilize the lesion and play a role in prevention of progression of the disease in adults [17,18].Radiation therapy is contraindicated because of a reported incidence of malignant degeneration with the formation of osteosarcoma[19].

Conclusion

Fibrous dysplasia is a benign tumour and therefore when it is asymptomatic it can usually be observed. Surgical intervention is indicated for symptomatic lesions causing intractable bone pain, severely reduced mobility, cosmetic deformity, or local pressure on vital structures. 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 cosmetic appearance. The best recommendation is to follow the wishes of the patient.

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