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Case report/Kazuistyka

An extensive Fibrous dysplasia of anterior skull base area of a 12-year-old boy – A case report

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ABSTRACT

Fibrous dysplasia (FD) is a relatively rare benign disorder of unknown etiology where normal bone is replaced by fibrous tissue with woven bony trabeculae. FD is an extremely rare bony tumor involving extensively skull base area in a child. FD has uncertain pathogenesis and diverse Histopathological pictures. Careful observation is needed as sometimes it become symptomatic which need immediate surgery in the head and neck region. We report a rare case of fibrous dysplasia involving extensively anterior skull base in a pediatric patient without presenting any clinical manifestation except swelling in right side facial area. As patient is asymptomatic, he is carefully monitored by us to treat earliest if symptoms appear.

Introduction

Fibrous dysplasia (FD) or Jaffe–Lichtenstein disease [1] is an uncommon benign bony tumor of unknown etiology where normal bony structure is replaced by fibrous connective tissue with woven bony trabeculae. FD is a rare benign tumor of bone and it accounts for 2.5% of all bony tumor and 7.5% benign tumors of skeletal system [2]. Von Rechlinghausen first described this characteristic lesion, called fibrous dysplasia in 1891 but Lichtenstein in 1938 introduced this term into worldwide literature [2]. There are two forms of FD, one is monostotic type which represents 70% of the cases and is characterized by only one focus of involvement [3]. The commonest sites for monostotic type are femur and costae. Polyostotic FD is seen with multiple foci affecting several bones. Sometimes the polyostotic type is associated with McCune Albright syndrome representing as an endocrine disorder and café-au-lait spots. Males and females are equally affected by fibrous dysplasia and often seen before the age of 30 years. Craniofacial involvement of fibrous dysplasia occurs in 25% of cases. Most common craniofacial bones are involved are maxillary and mandibular bone whereas the frontal, sphenoid and ethmoid are rarely involved [4]. The histological evaluation reveals a collagen matrix stroma with fibroblasts in an entangled standard with osseous trabeculate mimics to the "Chinese writing" [5]. Clinical symptom other than facial asymmetry may not be visualized in patient of FD. Blindness and deafness are two severe clinical symptoms in FD due to compressive

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manifestations. Here we are presenting a case of asymptomatic case of FD in 12 years old boy except a swelling in right side of face.

Case report

A 12-year-old boy attended the Outpatient department of Otorhinolaryngology with complaints of bulging of the right side facial area since 6 months (Fig. 1). He had no history of nasal obstruction, eye protrusion, visual disturbance and otalgia. He has no symptoms or signs of increased intracranial tension. His nasal examination was within normal limit. There was no family history of similar attack. The general physical examination showed a moderate built patient with normal vitals and no features of mental retardation. Intraorally there was no bulging in the oral cavity and oropharynx with healthy mucosa. Computed tomography (CT) scan of paranasal sinuses, orbit and brain showed that there was a heterogeneous lytic expansile lesion involving the right side facial bone, right sphenoid bone, right pterygoid plates and not involving orbit and brain (Fig. 2a & b). The mass in the CT scan resembles to the opaque glass appearance. Biopsy was taken sublabially, showed fibrous dysplasia. Histopathologically, it showed a overgrowth of nondescriptive fibrotic tissue with a secondary phenomenon of bony metaplasia (Fig. 3). As the lesion is asymptomatic, it was decided to keep the patient under observation.

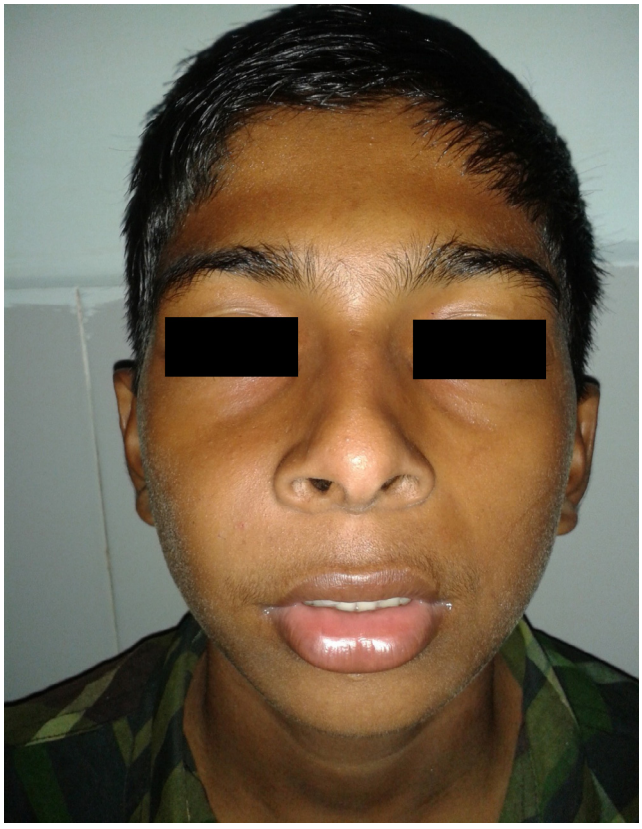

Fig. 1 – A 12 year old boy showing swelling at right side of face due to fibrous dysplasia

Discussion

FD is a benign lesion characterized by progressive replacement of bone with fibrous tissue. FD is a rare bony tumor occurring predominantly among young adults. Common locations of FD are thorax and extremities whereas it is rarely seen in head and neck area. There are four clinical types of FD have been seen. Monostotic FD which accounts for 70–80% FD and craniofacial region is involved in 10–25% of cases [4]. Here any one bony structure in craniofacial

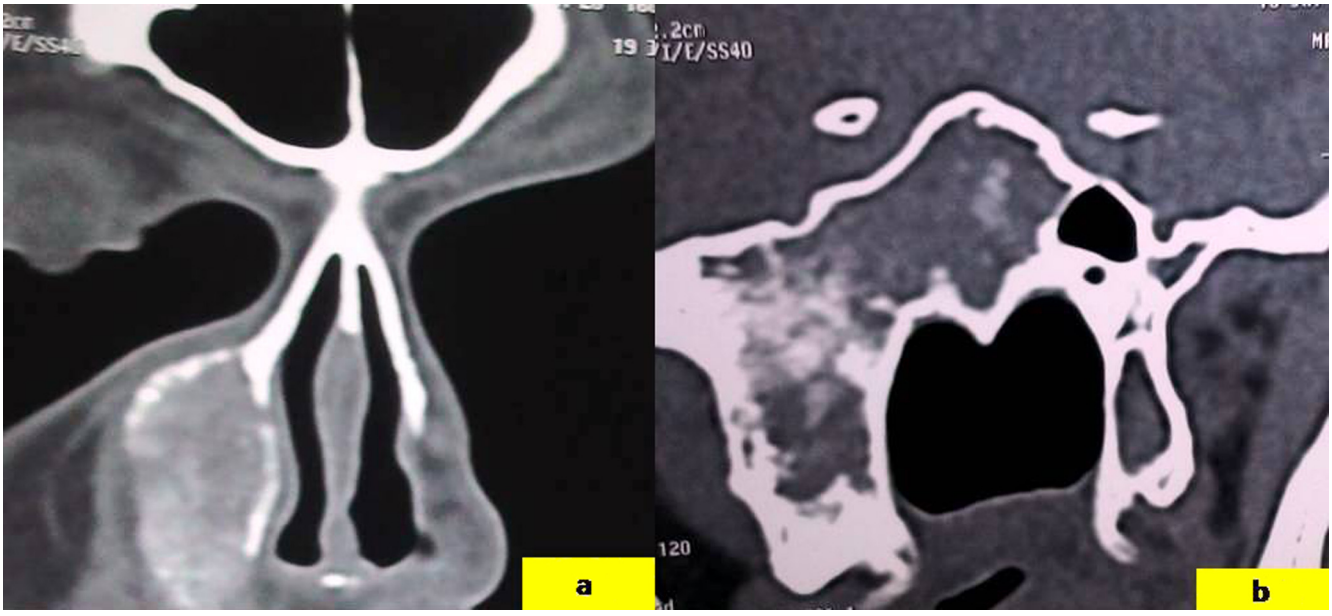

Fig. 2 – (a & b) CT scan of paranasal sinus showing tumor of heterogeneous mass involving frontal process of maxilla, maxillary sinus (a), sphenoid sinus, pterygoid plates (b)

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