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Case Report

Case Report of Diagnostic Dilemma – Fibrous Dysplasia/ Osteosarcoma

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ABSTRACT

Among bone tumours of head neck region, benign tumours of bone are common, while malignant tumours are very rare. These benign tumours constitute about 75% of all bone tumours. Most of these bone tumours present clinically with pain, swelling and symptoms of compression of vascular and neural structures. Surgery is not required for these bone tumours unless patient experiences any discomfort as diagnosis is made on plain radiographs. Only 2% of all benign tumours may undergo malignancy[1]. Fibrous dysplasia (FD) is a less common skeletal developmental anomaly of the bone, which is often misdiagnosed as malignant bone tumour. It manifests as defect in osteoblastic differentiation and maturation and affects a single bone or may involve multiple bones[2]. Osteosarcoma is a malignant mesenchymal tumour, predominantly affecting the long bones and occasionally seen affecting the maxillofacial region. It accounts for 15–35% of all primary bone tumours and 4–8% of sarcomas of jaw[4]. In osteosarcoma, males are more commonly affected than females. The aim of this article is to represent a rare case of bone tumour of the upper jaw in 10-year-old boy, which was initially suspected as osteosarcoma and later confirmed as FD.

KEYWORDS: Fibrous dysplasia, Maxilla, Young child, Osteosarcoma, Bone tumour, Diagnostic dilemma, Chinese letter pattern

CASE REPORT

A 10-year-old male patient was referred to the Oral pathology department to our college with a chief complaint of swelling in upper front tooth region since one month. Initially swelling was slow growing and attained the present size of 4×3 cm.

Extra-oral examination (Figure 1) revealed well-circumscribed round to oval shaped swelling present on upper left maxilla with no extra-oral swelling. Regional lymph nodes were not palpable.

Intra-oral examination revealed a well circumscribed solitary swelling seen extending from right canine to and left maxillary incisors, crossing midline on the labial and palatal side. The swelling measured 4×3 cm. On palpation, swelling was firm and non-tender. Radiographic examination revealed unilocular radiolucency.

Incisional biopsy was performed and the haematoxylin and eosin stained section showed a highly cellular connective tissue stroma, with numerous immature bony trabeculae arranged irregularly and osteocytes were seen present within the bony lacunae, which was suggestive of osteosarcoma (Figure 2). Areas of Osteoid were also seen with osteoclasts and (Figure 3,4). Numerous giant cells were also evident. Initially there was dilemma in the diagnosing the lesion, then the case was referred to higher centre for further evaluation. After referring the literature, it was finally diagnosed as fibrous dysplasia (FD).

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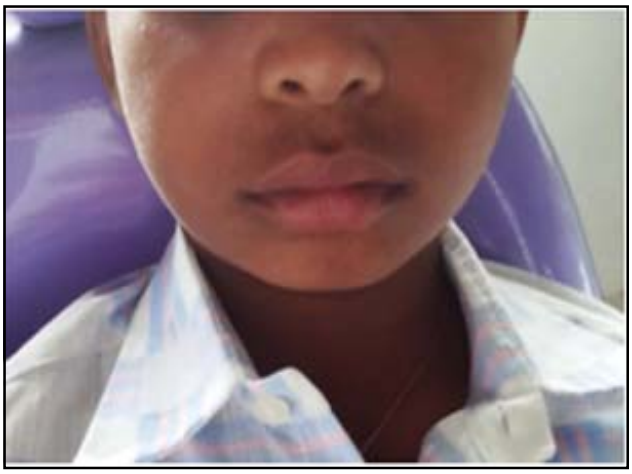


Figure 1: Extra oral view with no noticeable swelling

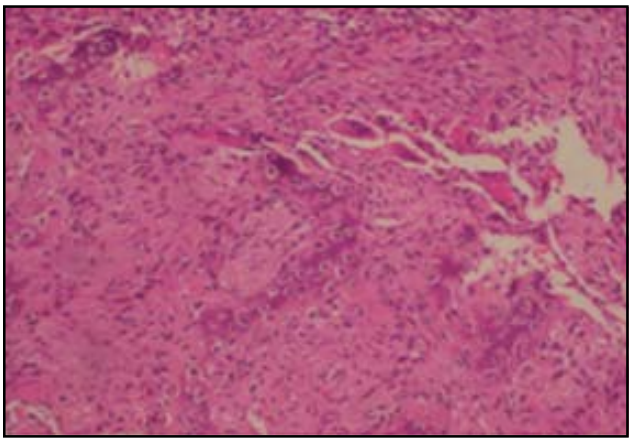
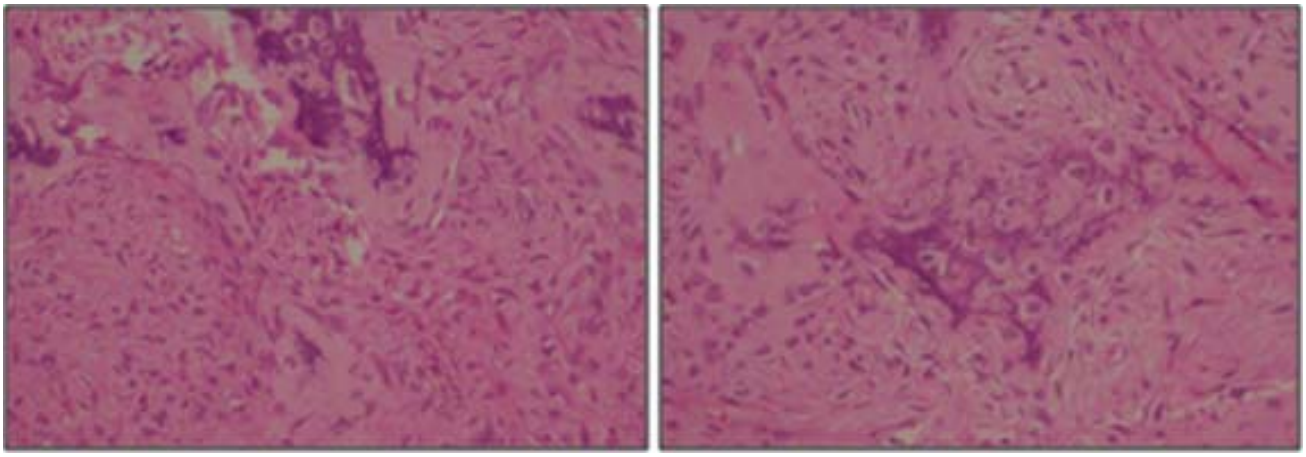


Figure 2: Fibrous connective tissue with irregular osteoid formation



Figures 3 and 4: Show fibrous connective tissue with areas of osteoclastic resorption

DISCUSSION

FD was first reported by Albright and his co-workers in 1937. The monostotic and polyostotic forms of FD were described by Lichtenstein and Jaffe in 1942. FD represents only 5–7% of benign bone lesions with involvement of the facial bones is much rarer. Gender prevalence of FD is equal[4]. The monostotic form of FD commonly affects the middle age group of 20–30 years age. Jawbones are more frequently affected in them than the extra-skeletal system. Children younger than 10 year of age are affected by polyostotic FD; lesion grows with the age and gets stabilised at puberty. Craniofacial bones, ribs, and diaphysis or metaphysis of the proximal femur and tibia are affected[4]. The ratio of occurrence of monostotic to polyostotic FD is 7:3. The etiology of FD has been explained by many theories, which signifies many etiological factors like congenital anomaly, trauma or any other non-specific disturbance in local bone reaction or 'perverted' activity of bone-forming cells and an endocrine disturbance with local bone susceptibility. Of these theories, Lichtenstein's theory of abnormal growth of undifferentiated osteogenic mesenchymal tissue is well accepted. The etiology of FD is now believed to be a

developmental error in which bony expansion may be due to primitive fibrous tissue proliferation within the bony medulla that encroaches upon the cortex[4]. Histopathologically shows irregularly shaped immature bony trabeculae in cellular, loosely arranged fibrous stroma. They often assume a curvilinear shape, resembling Chinese script writing. There is a monotonous arrangement of bony trabeculae rather than haphazard arrangement. Typical *Chinese letter pattern* of the trabeculae is characteristic feature. Lesional bone fuses directly to normal bone so that capsule or line of demarcation is not seen[5]. FD may be easily diagnosed, but some of its clinical and radiographic features mimic the other benign fibro-osseous lesions of the skeleton and rarely sometimes may be confused with certain types of malignancies like Osteosarcoma[5,6].

Osteosarcomas a primary malignant tumour of bone characterised by the production of immature woven bone from mesenchymal tumour cells. Usually develops at age of 15–25 year and site of involvement being metaphyseal plates of long tubular bones i.e., femur, tibia and humerus. Males are more frequently involved than females. In the head neck region, mandible is the most common site of involvement[7]. Histopathalogically lesion shows direct production of osteoid and chondroid material cells show variation in shape from relatively uniform round or spindle shaped to highly pleomorphic cells with bizarre nuclear and cytoplasmic shapes. Osteosarcomas should be considered in differential diagnosis of expansile lesions of jaws. Low-grade central osteosarcoma, a variant of osteosarcoma consists of fibroblastic stroma with variable degree of cellularity and with characteristic arrangement of osteoid in parallel seams[8]. Occasional mitotic figures with minimal cytological atypia are always present in low-grade osteosarcoma; whereas cytological atypia is not shown by FD which is an important distinguishing feature. Permeative growth pattern of FD is the most helpful factor in differentiating osteosarcoma, as the histological similarity is seen in both lesions; the radiographic features play an extremely important part of the diagnosis. Radiographic features of low grade osteosarcomas are variable most, commonly seen pattern is a large expansile intra-compartmental fibro-osseous lesion with coarsely thick orthin incomplete trabeculations, an another type of pattern is a dense sclerotic lesion showing cortical erosion and soft tissue extension.

The key feature to distinguish LGCO from the benign lesions of jaws is to identify its permeative growth pattern. Diagnosis is difficult on biopsy samples particularly when only the fibroblastic stroma devoid of cytologic atypia is represented and when permeation of the host trabecular bone cannot be demonstrated. Radiologically FD, desmoplastic fibroma, non-ossifying fibroma, osteoblastoma, and aneurysmal bone cyst may be confused with LGCO, it is therefore vital to combine both radiological and histological findings to give an accurate diagnosis of FD[8].

CONCLUSION

FD is a rare case of head and neck region. OS should always be considered in the differential diagnosis of expansile lesions of the jaws. As clinical features of FD overlap with osteosarcoma, careful clinical, radiographic and histological findings must be carried out to confirm the diagnosis of FD.

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