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AN APPARENTLY AGGRESSIVE CRANIOFACIAL OSTEO-DYSPLASTIC LESION PRECIPITATING DEBILITATING SYMPTOMS AND SIGNS: CASE REPORT
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# AN APPARENTLY AGGRESSIVE CRANIOFACIAL OSTEO-DYSPLASTIC LESION PRECIPITATING DEBILITATING SYMPTOMS AND SIGNS: CASE REPORT

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## SUMMARY

**This is a case report of an 18 year old man with craniofacial fibro-osteo-dysplastic lesion which exhibited both exophytic and endophytic growth patterns. We discuss the extent of tumour growth and its associated secondary changes.**

## INTRODUCTION

Fibro-osseous lesions (FOLs)of the craniofacial complex represent a heterogeneous group of lesions characterised by replacement of normal bone with collagen and fibroblasts with varying degree of mineralised matrix containing bone or cementum (1). FOLs are mostly composed of benign tumours such as fibrous dysplasia (FD), cemento-osseous dysplasia (COD) and ossifying fibroma (OF) or cemento ossifying fibroma (COF). Malignant degeneration occurs in less than 1% of the cases. Malignancies are almost exclusively osteosarcoma and Ewing's sarcoma (2). Because of significant histopathological similarities among the FOLs, the definitive diagnosis rests on the constellation of clinical, radiological and histologic findings. This case report describes a rare and peculiar form of craniofacial FOL. It shows the geographical extent of the tumour as well as its associated secondary changes.

## CASE REPORT

An 18 year old slightly disoriented boy presented at M.P.SHAH hospital (CT Department) with a history of progressive facial swelling, headache and proptosis of the right eye.Patient had no prior history of surgery or radiotherapy. Pre-contrast and post-contrast CT scans were done which showed an expansile complex bifrontal mass of osseous density exhibiting both exophytic and endophytic growth patterns.The mass was largely sclerotic with randomly distributed areas of hypointense foci.Endophytically, the lesion showed an extension to the anterior cranial fossa (Figure 1). This contributed to the mass effect on the adjacent cerebral cortex with perilesional oedema. A bony island with ill-defined edges and a central hypointense centre was also noted (Figure 2). There was also bilateral effacement of the cortical sulcal spaces,sylvian fissures and basal cisterns indicating thepresence of araised intracranial pressure. However, no midline shift was evident or abnormal enhancement after intravenous contrast media.Exophytically, there was extensive destruction and obliteration of the frontal bone. Expansile growth of the osseous mass was beyond the cortical confines of the frontal bone (Figure 3).Inferiorly, it extended to the right orbital roof causing compression and inferior displacement of the orbital contents (Figure 4). There was significant diminution of orbital volume but no direct invasion seen.

**Figure 1**
*Post-contrast axial reformat*

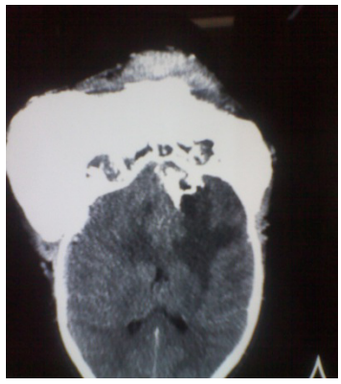


**Figure 2**
*Post-contrast coronal*

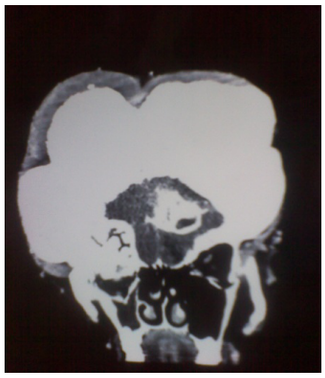
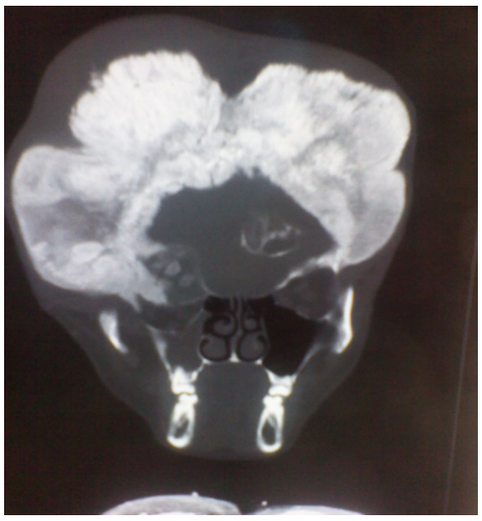


**Figure 3**
*3-D Reconstruction*

**Figure 4**
*Bony window*



## DISCUSSION

Fibro-osseous lesions (FOL) of the craniofacial complex comprise a diverse and a challenging group of conditions that pose difficulty in classification and diagnosis (3).The current classification includes neoplasms, developmental dysplastic lesions and inflammatory / reactive processes (4). They are mostly composed of benign tumours such as fibrous dysplasia (FD), cemento-osseous dysplasia (COD) and ossifying fibroma (OF) or cemento ossifying fibroma (COF) (5). Although, there is no widely accepted agreement, FD,CD,OS,are considered to be developmental or reactive regenerative abnormal proliferation. However, COF is a true neoplasm, benign in nature (6). Ossifying fibromas (OF) and Fibrous Dysplasia (FD) are the most common benign FOLs (3).

FD is a developmental dysplastic disorder of bone in which normal bone matrix is replaced by fibroblastic proliferation. Craniofacial FD is uncommon and accounts for 10% of all monostotic forms of the disease and nearly 100% of polystotic with the mandible and the maxilla being the most common affected sites. Involvement of the ethmoid, sphenoid, frontal and temporal bones are infrequent. Extensive FD of the frontoethmoisphenoid region is uncommon but is more aggressive (7). Critical to the diagnosis is the fact that FD fails to manifest any discrete margins; rather the lesional bone subtly blends into the surrounding normal appearing bone (4). Intracerebral extension of the tumour is a rare occurrence. FD has characteristic appearances on CT and consists of three varieties: ground glass pattern (56%), homogenously dense pattern (23%) and cystic variety (21%) (3). Although these are typical features, it should be noted there are other fibro-osseous conditions that may mimic FD such as osteoma,chordoma,hyperostotic meningioma, aneurysmal bone cyst and Pagets disease.

Ossifying fibromas (OF) and cemento ossifying fibroma (COF) are regarded as a single lesion of fibro-osseous entity. There is only a difference in histopathological findings on bone between calcified element and cementum (8). Though commonly seen in the mandibles in females, it can arise in the paranasal sinuses and skull base, behaving in a more aggressive manner at the intracranial and orbital interfaces (1). OF are associated with a slowly progressing enlargement of the affected bone. Death is a rare occurrence secondary to intracranial extension. CT findings can be fairlydestructive, circumscribed lesions with sclerotic eggshell rim and central radiolucency with a variable amount of internal calcification (1).Cemento-osseous dysplasia (COD) is a group of non-neoplastic processes usually confined to the tooth bearing areas of the jaws or edentulous alveolar processes. They consist of periapical, focal, and florid cemento-osseous dysplasia (9). Most malignant FOLs are osteosarcoma

and Ewing's sarcoma.Osteosarcoma is a malignant tumour caused by the transformation of mesenchymal cells which have capacity to generate osteoid tissue and mature woven bone. They are characterised by orthoiradial striations, destruction of the cortices with an outgrowth of soft tissue component (10).

In conclusion, FOLs have similar features with each other. However, such similarity induces difficulties in differential diagnosis. It's crucial to isolate malignant lesion with benign ones (7). CT is a better radiological tool, especially for assessing the extent of the tumour. However, CT alone is insufficient to make a diagnosis (7). Precise clinicoradiologic and / or histopathological diagnosis is imperative as the management can vary significantly for these lesions. This can range from serial observation to aggressive surgical resection based on symptomatology, size and location of the FOL (1).

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