A 49-year-old female was operated for left lateral orbital wall fibrous dysplasia 12 years back. The patient was having a relatively uneventful course till four months back when she started to have headache and noticed a swelling in the scalp which gradually increased in size over a period of time. On examination she had a mild proptosis of the left eye with a normal vision. There were two irregular bony swellings in the left side of the head. Patient had no other symptoms. Bony windows of noncontrast CT head showed an expansile mass lesion in the left orbito-fronto-temporal region with sclerotic rim with a bony defect. A similar swelling was noted in the parietal region, suggestive of fibrous dysplasia [Figures 1 and 2]. The intervening bone between the two lesions was absolutely normal. Surgery was performed and shaving of both the orbito-fronto-temporal and parietal bony lesions with cranioplasty was done. Orbital decompression was done along with it to preserve the patient’s vision and to correct the proptosis. Intraoperatively, the findings were consistent with a diagnosis of fibrous dysplasia which was confirmed on histopathology. The headache improved postoperatively. Postoperative reconstructed 3D skull image showed the repaired lesions separated by normal intervening bone exquisitely well [Figure 3].

Fibrous dysplasia is a bone disorder of unknown origin characterized by slow, progressive replacement of bone by abnormal proliferative isomorphic fibrous tissue.[1-3] The disease was first described by McCune and Bruch in 1937.[2] Most lesions are monostotic, asymptomatic and identified incidentally which can be treated with clinical observation and patient education.[3] Calvarial involvement is usually monostotic and is seen in only 10% of cases.[2]
Sphenoid, frontal, ethmoid and maxillary bones are affected in the order of frequency.\(^4\) Involvement of multiple sites in the calvarium, spatially separated by areas of normal bone as described in this case.

It is generally believed that the condition is more frequent in children and does not increase after the period of bony growth. Rarely, the disease is also reported in adults.\(^5\) More often the patients present with headache and facial asymmetry, due to the enlargement of the facial bones. Painless proptosis is not uncommon, being mild in most of the cases.\(^5\) Visual dysfunction and optic atrophy occur in lesions involving the sphenoid wing, encroaching upon the optic canal.\(^5\) Three forms, pagetoid, sclerotic and cystic are described with this condition, with imaging generally revealing a classic ground glass appearance of the sclerotic bone on CT.\(^4\) Surgery is indicated for confirmatory biopsy, to correct cosmetic deformity incurred due to overgrowth of bone or decompression of the neural structure producing symptoms,\(^4,5\) as these lesions are resistant to radiotherapy and chemotherapy.

### References


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