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Fibrous dysplasia of sphenoid bone
presenting as a case of loss of vision

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ABSTRACT

Background: Fibrous dysplasia is a fibro-osseous lesion of unclear aetiology wherein normal bone is replaced by abnormal fibrous tissue and immature bone. Fibrous dysplasia is associated with a defect in osteoblastic differentiation and maturation that originates in the mesenchymal precursor of the bone & is well documented to affect craniofacial structures.

Case description: A case of the lesser sphenoid wing fibrous dysplasia is described which presented with symptoms of pressure effects on the optic nerve, managed subsequently with microsurgical decompression of the nerve.

Conclusion: Craniofacial fibrous dysplasia is an uncommon entity which can present with loss of vision, wherein the visual prognosis depends upon timed & adequate surgical intervention.

INTRODUCTION

Fibrous dysplasia is associated with a defect in osteoblastic differentiation and maturation where normal medullary bone is replaced with a variable amount of abnormal and structurally weak fibrous and osseous tissue. It is of particular interest to neuro-ophthalmologist as it can affect craniofacial bones wherein encroachment on the paranasal sinuses, orbit, and foramina of the skull can produce variety of ophthalmological symptoms. Here in a case is described of fibrous dysplasia of lesser sphenoid wing presenting with optic nerve compression & visual loss.

CASE REPORT

A 45-year-old female housewife presented with holocranial headache for 1 year which was dull aching, moderate intensity, and gradually progressive, non-pulsatile, no specific aggravating factors, relieved on medication. There was no history of vomiting, seizures, trauma, aura and photophobia, facial pain & anosmia. She also complained of

Keywords

fibrous dysplasia,
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blurring of vision followed by gradual progressive painless, visual deterioration in right eye for 2 months. There was no history of painful movements of eyeball, reddening/ congestion, diplopia, proptosis, epiphora/ glare. She didn't complain of tunnelling of vision & had no history suggestive of hormonal changes.

Examination revealed the visual acuity in right eye as 6/60 & left eye 6/12 unchanged with pinhole, pupillary response was normal in left eye whereas relative afferent pupillary defect was present in right eye. On perimetry, paracentral scotoma was observed on right side while left side was normal. Fundus examination, extraocular movements & conjunctiva were normal on both sides. On CT scan of brain, orbit & skull base, diffuse expansion of medullary cavity with ground glass bone matrix with no contrast enhancement & intact cortices was noted involving bilateral greater & lesser wing of sphenoid, bilateral anterior clinoid processes & pterygoid plates with orbital wall & roof thickening on both sides causing narrowing of bilateral optic canals (right more than left) (Fig.1). Blood investigations were normal.



Figure 1.

Based on imaging findings, a provisional diagnosis of fibrous dysplasia was made. Right frontal craniotomy was performed and optic nerve decompression was achieved by drilling of right anterior clinoid process and orbital roof. Intra-operatively right sphenoid wing, frontal bone, anterior clinoid process and orbital roof was found to be thickened and bone was spongy and vascular while the optic nerve seemed to be compressed in optic canal, with thinning of intracanalicular portion. Optic nerve decompression was done till orbital apex (Fig 2a & 2b).

Histopathological examination revealed irregular trabeculae of woven bone within fibrous tissue which lacked osteoblastic rimming suggestive of fibrous dysplasia. Post operative CT scan showed significant drilled out bony portion at right orbital roof thus demonstrating decompression of the optic

nerve as compared to preoperative scans (Fig 3 & 4). Visual acuity in right eye as assessed at the end of 1 month & 2 months of follow up period was 6/36 & 6/24 respectively, hence showing betterment; along with improvement in headache.

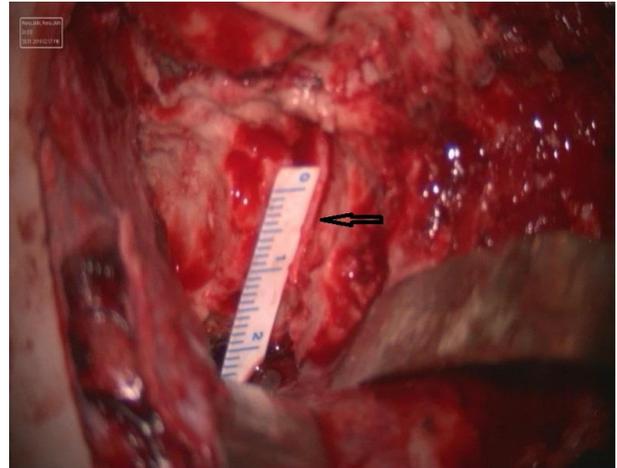


Figure 2a.

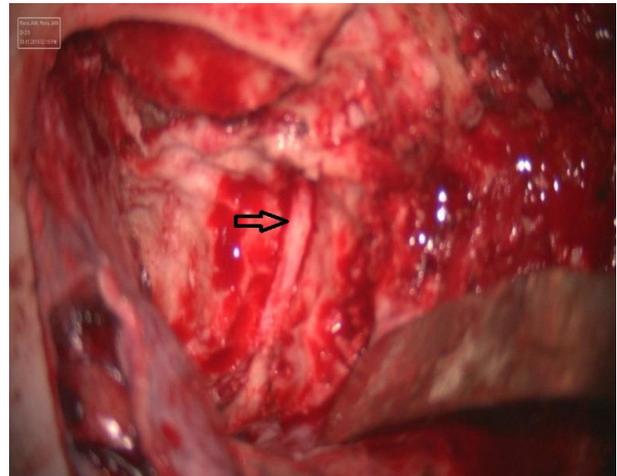


Figure 2b.

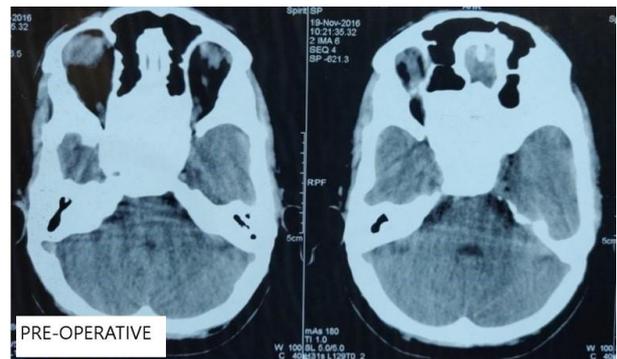


Figure 3.

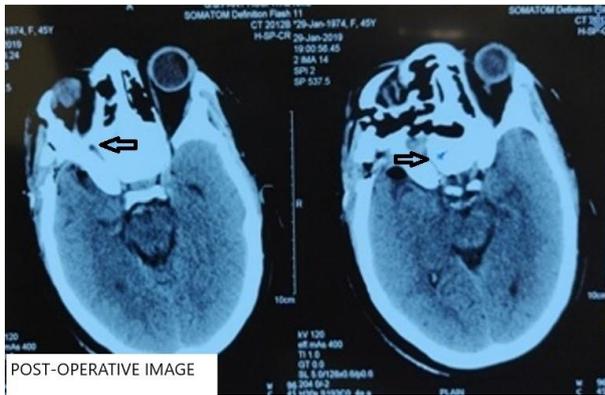


Figure 4.

DISCUSSION

Fibrous dysplasia was first recognized by von Recklinghausen in 1891 and further characterized and named by Lichtenstein in 1938. It is one of the most common benign skeletal disorders and can affect any bone in the body [2]. It is most common in adolescents and young adults, and progression decreases after puberty [2]. There are three types of fibrous dysplasia: monostotic (the most common, accounting for 70% of cases), polyostotic, and McCune-Albright syndrome. McCune-Albright syndrome is characterized by polyostotic fibrous dysplasia, endocrine hyperfunction (which can lead to precocious puberty in female patients), café au lait pigmentation of the skin, and other extraskeletal abnormalities [1].

Fibrous dysplasia has a predilection for the skull base; particularly the ethmoid (most common) followed by sphenoid, frontal, maxilla bones & orbits [5]. Consequently, patients with fibrous dysplasia may exhibit local cosmetic deformity (the most common manifestation), headaches, proptosis, impaired ocular movement, and loss of visual acuity [2].

Based on radiographic features, fibrous dysplasia is classified into 3 types, the pagetoid, or “ground-glass,” pattern is the most common (56% of all cases); it appears as a mixture of dense and radiolucent areas of fibrosis. The sclerotic pattern (23% of cases) is uniformly dense. The cystic pattern (21%) is characterized by an spherical or ovoid lucidity surrounded by a dense bony shell.[3]

Involvement of the orbit leading to visual loss is the most feared complication, and surgical decompression may be warranted in acutely or progressively symptomatic patients (or in both) [1,2]. Radiotherapy and chemotherapy have no role in the treatment of fibrous dysplasia, and the former may increase the risk for malignant degeneration (most commonly osteosarcoma) [3,4].

Fibrous dysplasia may be complicated by aneurysmal bone cyst formation [1]. Malignant degeneration occurs in 4% of cases of fibrous dysplasia and is more common with the monostotic type. The interval between the diagnosis of fibrous dysplasia and evidence of sarcomatous degeneration is long, with a mean of 15 years. Close imaging follow-up with serial CT and MRI is essential in patients with known fibrous dysplasia. Worsening pain, development of a soft tissue mass, or elevation of alkaline phosphatase levels should raise concern for malignant degeneration [2].

CONCLUSION

Fibrous dysplasia of the sphenoid bone, although not a common entity, can present with symptoms of visual loss. Timely intervention & adequate microsurgical decompression of the optic nerve results in good postoperative visual outcome and is of prognostic significance.

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