



Sudden Bilateral Vision Loss Due to Ruptured Sphenoid Mucocele in Fibrous Dysplasia: A Case Report

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ABSTRACT

Fibrous dysplasia is a developmental anomaly of bone, often affecting the facial bones. We present a case of fibrous dysplasia with bilateral sudden loss of vision. MRI showed ruptured sphenoid sinus mucocele compressing the optic nerves. Immediate surgical exploration and decompression restored the vision.

KEYWORDS

Fibrous dysplasia, Mucocele, Vision Loss, Optic Nerve Decompression.

Introduction

Fibrous dysplasia is a developmental anomaly of bones, often affecting the facial bones [1]. It is caused by a post-zygotic, activating mutation of the GNAS gene leading to replacement of normal marrow and bone by fibrous tissue and woven bone [2]. Involvement of the craniofacial bones can produce visual loss, proptosis, diplopia and epiphora [3]. Mucoceles are benign cyst like lesions lined with respiratory epithelium and encapsulated masses that may lead to destruction of bony structure surrounding it [4]. Most prevalent location of sinus mucocele is frontal sinus and only 1% occurs in sphenoid sinus [5,6,7]. Sudden vision loss by sphenoid mucocele is rare. So herein we present a case of sudden onset bilateral vision loss in a patient of fibrous dysplasia caused by sphenoid sinus mucocele in which immediate decompression of the optic nerve restored the vision.

Case Report

A 32 year old male was brought to the hospital with history of headache from three days and bilateral vision loss from last 4 hours. There was no complaint related to vision in the past. General physical examination revealed an asymmetric skull deformity with a bony swelling over the right frontal bone. There was 7 cm shortening of right lower limb. Visual acuity of perception of light in right eye and finger counting at 1 metre in left eye was found. Fundus examination revealed minimal temporal pallor of optic discs bilaterally. CT head and MRI brain were done besides the routine investigations. X ray Pelvis showed ground glass matrix with bowing deformity of right hip bone (Figure 1) and CT head revealed expansile calvarial lesion more on right side of frontal bone, right orbital plate, right parietal bone and occipital bone with features consistent with polyostotic fibrous dysplasia (Figure 2). MRI revealed sphenoid sinus mucocele extending into the clivus with air-fluid level suggestive of ruptured mucocele of sphenoid sinus (Figures 3a & 3b). A diagnosis of ruptured sphenoid sinus mucocele was made and immediate decompression was done

on the same day via transthemoid approach. Patient gradually regained the vision with visual acuity of 6/9 bilateral eyes two weeks after the surgery.

Discussion

Craniofacial fibrous dysplasia associated with acute vision loss has been reported with mucoceles, hemorrhage and hemorrhagic cysts, as well as with fibrous dysplasia alone when the optic canal is involved [3]. Several cases of sudden acute loss of vision caused by sphenoid mucocele have been reported in the literature [8,9,10]. Weissman JS et al (1990) reported a case of fibrous dysplasia with sudden loss of vision. Radiological investigations revealed mucocele at the orbital apex and optic canal compromise by bony proliferation. Surgical excision of the cyst and debulking of the fibrous dysplasia improved the vision [1]. Dowler JG et al (1995) reported a case of sudden bilateral loss of vision in Albright's syndrome which was treated with surgical decompression with resultant improvement in vision. Patient got vision of 6/9 in each eye one year after surgery [9]. Papadopoulos MC et al (1998) reported two cases of acute reversible visual loss in which one patient had a sphenoid sinus mucocele compressing the optic chiasma and the other had optic nerve narrowed by dysplastic bone. In both cases optic nerve decompression restored vision to normal [10]. In our case it was the prompt diagnosis and immediate surgical decompression which resulted in regaining the vision loss.

Conclusion

Prompt diagnosis and immediate surgical decompression can be sight saving in cases of sphenoid mucocele associated with vision loss.



Figure 1. Image shows homogeneous loss of the normal trabecular pattern in the shaft of the right femur, with a ground-glass appearance caused by fibrous dysplasia.

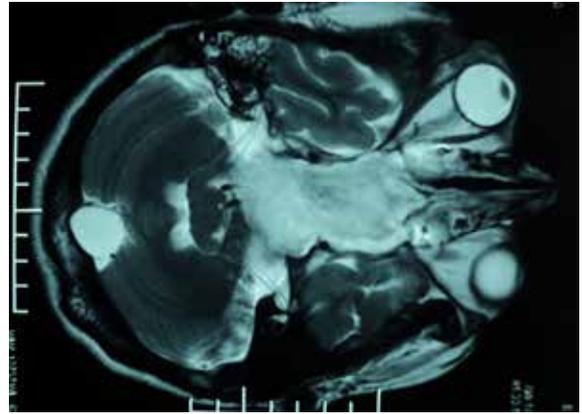


Figure 3b: Axial MRI T2W1 image showing ruptured sphenoid mucocele compressing optic nerves.

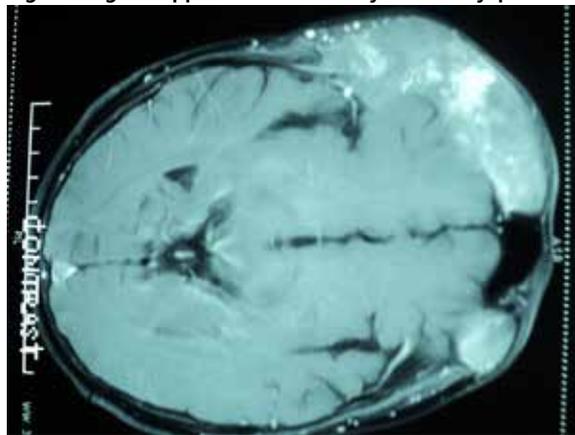


Figure 2. Axial CT scan shows a expansile bony mass of frontal and parietal bones ; this finding is consistent with fibrous dysplasia. Note the relative homogeneous attenuation of the lesion.

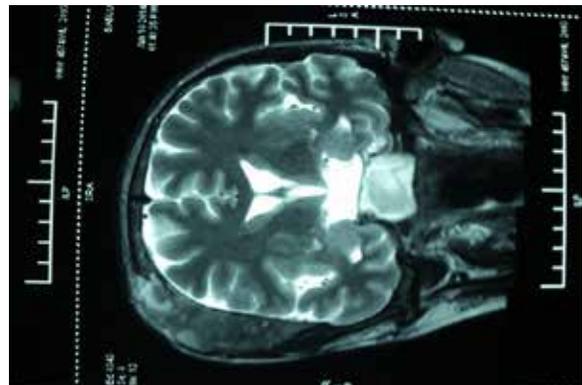


Figure 3 a : MRI T2W1 coronal section showing ruptured sphenoid sinus mucocele and right parietal bone expansile mass caused by fibrous dysplasia.

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