Case report

Anterior clinoid mucocele causing optic neuropathy: A case report and review of literature

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ARTICLE INFO

Keywords:
Anterior clinoid process
Mucocele
Optic neuropathy

ABSTRACT

A 66 year old Indian gentleman presented with a 3 days history of headache and gradual progressive loss of vision in his left eye, ophthalmological assessment showed no light perception in his left eye with papilledema and afferent papillary defect. Computed tomography (CT) and Magnetic Resonance Imaging (MRI) were done and showed an expanding lesion in the left anterior clinoid process encroaching upon the left orbital apex and optic nerve with features suggestive of a mucocele. Patient was started on dexamethasone, and urgent craniotomy was undertaken, where marsupialization and resection of left anterior clinoid mucocele was done, and histopathologic examination of the operative specimen was consistent with a mucocele. Post-operatively, patient was kept on dexamethasone for few days, with uneventful outcome, and his follow up at 6 months showed complete recovery of his vision from no light perception to 6/12 in the affected eye.

1. Introduction

The anterior clinoid process (ACP) is a bony structure on the superolateral aspect of the sphenoid sinus lying between the internal carotid artery and the optic nerve. Pneumatization of the sphenoid sinus, can variably extend into different parts of the sphenoid bones, including the ACP, extension of pneumatization of the ACP with a frequency of around 9.5% [1,2,3].

Blockage of the drainage of a paranasal sinus for any reason can result in accumulation of trapped secretions and result in a mucocele. Mucocele of the ACP can result in compression on the adjacent structures in the optic nerve canal or superior orbital fissure, causing optic neuropathy (compressive or inflammatory) or ophthalmoplegia with dysfunction of third, fourth and/or sixth cranial nerves [4,5,6]. CT and MRI are complementary for the diagnosis, displaying bony anatomy, and related soft tissue changes and surgical treatment is usually advocated as soon as possible to achieve best recovery. Surgical options include open surgical resection or endoscopic approach along with medical treatment with steroids and antibiotics [6]. We are presenting a case of anterior clinoid ACP mucocele as a rare cause of optic neuropathy and complete visual loss which recovered fully after surgical decompression. To our knowledge, there are only 18 cases of anterior clinoid mucocele causing optic neuropathy or ophthalmoplegia reported in the literature (with no language barrier).

2. Case report

A 66-year-old Indian gentleman presented with headache and gradual painful loss of vision in the left eye that started 3 days back that rapidly progressed to complete loss of vision in his left eye. Ophthalmological assessment showed no light perception in the left eye with papilledema and afferent papillary defect, with normal extraocular movements and normal vision in the right eye.

CT of the skull base and brain showed pneumatization of ACP bilaterally with soft tissue density lesion measuring about 10 × 16 × 9 mm expanding the left anterior clinoid process causing compression left orbital apex and left optic nerve suggestive of mucocele (Fig. 1).

MRI showed area of abnormal signal intensity within the expanded left ACP with hyperintense signal on T1-W images and intermediate signal on T2-W images, no suppression of the T1 hyperintense signal on fat saturation images indicating high protein content, and marginal mucosal lining post-I.V. gadolinium contrast enhancement (Fig. 2).

The patient was started on dexamethasone, and was then taken for surgery.
surgery, where a left pterional craniotomy was done and through extradural approach the expanding ACP was drilled away allowing exposure of a greyish white soft lesion morphologically consistent with mucocele; the lesion was resected and sent for histopathology, the roof of the optic nerve canal was also drilled decompressing the optic nerve, and the surgical defect was filled with autologous fat graft (Fig. 3). In addition, histopathology result confirmed the diagnosis of mucocele.

The post-operative course was uncomplicated and patient tolerated surgery well. He was kept on dexamethasone for 5 days. The retro-orbital pain improved immediately post-operatively, his vision showed gradual improved to complete recovery on his 6 month follow up. Follow up MRI showed complete resolution of the anterior clinoid mucocele with no residual abnormality.

3. Discussion

Multiple theories postulated for development of paranasal sinus mucocele, with obstruction of the sinus ostium as a result of mucosal thickening, fibrosis, bony overgrowth or tumors causing retention of mucous with erosion and remodeling of the adjacent bones [7]. It usually involves the frontal or the ethmoid sinus and rarely the sphenoid sinus [13], and involvement of the clinoid process as a localized lesion without sphenoid sinus involvement is very rare and causing symptoms is even more rare [14].

As described by Lim et al. [8], pneumatization of the anterior clinoid process can occur as part of the normal development of the

Fig. 1. Preoperative CT axial cuts at the level of the orbit and skull base, a. bone window display, and b. soft tissue window display, showing the expanding mucocele as soft tissue density, with thinning out of remodeled left ACL (arrow in a, and b).
sphenoid sinus, most often bilaterally, lined by respiratory epithelium with the potential of developing mucocele if its drainage is blocked. Chung et al. and Osborn et al. proposed a different theory and believed that ectopic mucosal cells trapped during embryogenesis can be due to direct compression on the cranial nerves causing ischemia within the developing bone cause mucocele formation [15,16].

ACP mucocele can expand causing bony enlargement and remodeling and due to the proximity to optic nerve and the superior orbital fissure, it can present with a variety of symptoms including retro orbital pain, blurring of vision, monocular visual loss, with variable degree of visual field defect, diplopia due to compression 3rd, 4th or 6th cranial nerves, and sensory deficits of trigeminal nerve [9,12,21], symptoms can be due to direct compression on the cranial nerves causing ischemia and dysfunction, or possibly due to extension of the inflammatory process from the mucocele to the nerve fibers causing neuritis.

Rapidly deteriorating vision is a bad prognostic sign, and urgent imaging and management should be undertaken immediately, and combination of retro-orbital pain and unilateral visual loss should not be superficially considered as retro bulbar neuritis without excluding a compressive etiology. CT is usually the first screening line to rule out compressive lesion in a patient with visual loss or diplopia, and should be followed by MRI to better visualize the related neurovascular structures. ACP mucocele usually appears as a rounded homogenous expansible lesion with surrounding bony morphology varying from thinning up to being eroded or absent in some areas [17,18,19]. The ACP mucocele may protrude into the sphenoid sinus or appear as dumbbell — shaped mass located between the optic nerve and the carotid artery [20]. MRI gives better visualization of the lesion, its contents, and the surrounding neurovascular structures. The signal intensity of the mucocele varies depending on the protein and water content composition, which varies with temporal evolution; becoming more hyper intense on T1 and hypointense on T2 with increasing chronicity as the protein content goes up and less hyper intense on T2 W images as the water content decrease. Thus, many possible combinations of signal intensities on MR images may be seen with mucocele, depending on their stage of development, water and protein content [22]. They do not enhance except for peripheral rim mucosal lining enhancement on post-I.V. gadolinium scans [6,8]. On MRI, the appearances may raise differential consideration of aneurysmal bone cyst, meningioma and schwannoma.

The severity and duration of symptoms affect the prognosis, and recovery of the patients. The treatment options vary from medical management with steroids and antibiotics to surgical decompression and marsupialization of the mucocele which can be done as an open procedure or endoscopically [4,6,8,9,10,11].

Medical management alone is often not sufficient in cases with neurological deficit, and surgical decompression is the main stay in such instances, and should be done as soon as possible, as timely decompression is the most important prognostic factor [6,9,10,11].

4. Conclusion

We herein report a unique case of anterior clinoid mucocele presenting with acute visual loss, which recovered dramatically after urgent surgical decompression. The timely radiological diagnosis and immediate surgical decompression was crucial to achieve for full recovery of vision.

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