Photo Essay

Sphenoidal mucocele presenting as acute cranial nerve palsies

Clarissa S.M. Cheng, MBBS, MMED(Ophth)(S’pore) a; Srinivasan Sanjay, MBBS, MRCS(Edin)(Ophth), MMED(Ophth)(S’pore), MS(Ophth), DNB(Ophth), MNAMS h,c; Chee Chew Yip, MBBS, MMED(Ophth)(S’pore), FRCS(Edin), FAMS a,b; Heng-Wai Yuen, MBBS, MRCSE, Mmed(ORL), DOHNS(Eng) d

Abstract

Sphenoidal sinus mucoceles are indolent lesions that, when sufficiently large, can compress on the optic canal or superior orbital fissure, rapidly causing loss of vision, optic neuropathy, ptosis, pain, ophthalmoplegia, and diplopia. We herein report a 72-year-old gentleman who presented acutely with Cranial Nerve II, III, and IV palsies secondary to a sphenoidal sinus mucocele that was confirmed on magnetic resonance imaging and successfully treated with endoscopic drainage. This cause of orbital apex syndrome is important for clinicians to know as early diagnosis and treatment is critical in recovering visual potential.

Keywords: Sphenoidal mucocele, Acute cranial nerve palsies, CT scan, MRI scan, Endoscopic sinus surgery

Case history

A 72-year-old Chinese male with a history of hyperlipidaemia and hypertension presented with a 10-day progressive droop of the right upper lid. Ophthalmic examination revealed Snellen’s visual acuity of 6/15 in the right eye and 6/9 in the left. There was a right partial ptosis and grade 2 relative afferent pupillary defect (RAPD). Extra-ocular motility (EOM) was limited in all directions except abduction (Fig. 1) and the colour vision was impaired. Confrontation visual fields, fundus examination, corneal sensation, and the remaining cranial nerve (CN) examination were normal. Ophthalmic examination of the left eye was normal.

Clinical findings were consistent with those of a combined CN II, III and IV palsy; hence the differential diagnoses of orbital apex syndrome was considered and urgent magnetic resonance imaging (MRI) was performed.

MRI brain and orbits showed a large hyperintense cystic lesion on T1-weighted images consistent with a sphenoidal mucocele. The mucocele expanded the right sphenoid sinus, extending to bilateral posterior ethmoid air cells, and compressing on the right superior and inferior orbital fissures (Fig. 2). The patient was immediately referred to the otolaryngologist who drained the mucocele endoscopically.

At one-week follow-up, there was a marked clinical improvement of visual acuity (6/9), ptosis, and colour vision. The pupil had a grade 1 RAPD and EOM was now partially limited to the up- and down-gaze (Fig. 1).

Discussion

Paranasal sinus mucoceles are slow growing benign cystic lesions containing mucoid secretions that can lead to sinus expansion, bony erosion, and extension into the orbit, cranial cavity and nasopharynx. They represent approximately 8% of all orbital tumours, with the frontal sinus mucoceles being the most common (50%), followed by frontoethmoidal (31%), ethmoidal (16%) and sphenoidal (3%). 1 They are caused...
by a blockage of the sinus cell ostium which can occur spontaneously or result from infection, chronic inflammation, trauma, iatrogenic injury, or tumour growth.2

The clinical manifestation of sinus mucoceles is dependent on the extent of the mass effect produced. Frontoethmoidal and maxillary sinus mucoceles tend to manifest as disorders of the globe and orbit due to their close anatomical association and this includes proptosis, enophthalmos and superior or inferior globe displacement.2,3

In contrast, the sphenoidal sinus is closely related to CN II–VI, pituitary gland, cavernous sinus and internal carotid artery, with only a thin lamina of the bone – lamina papyracea – separating them. An enlarging mucocele can easily disrupt this precarious anatomic relationship, with subsequent displacement and compression of these vital structures leading to the development of corresponding clinical syndromes. Should the sphenoidal mucocele extend to and impinge on the optic canal or superior orbital fissure, loss of vision, optic neuropathy, ptosis, pain, ophthalmoplegia, and diplopia supervene.4 Thus, although benign, when sufficiently large, sphenoidal mucoceles are sight-threatening and can lead to permanent visual loss.

Computed tomography (CT) and MRI are the imaging techniques of choice. Characteristic CT findings show a hypodense, nonenhancing mass that fills and distorts the sinus cavity. On MRI, the appearance varies according to the protein concentration of the mucoid secretion.5 Hyperintensity on T1-weighted signals indicated proteinaceous or mucoid material. The definitive treatment of choice for most mucoceles is intranasal drainage and marsupialisation using endoscopic sinus surgery (ESS).3

As illustrated in our patient, he presented with acute ptosis and associated CN palsies involving the optic, oculomotor and trochlear nerves. MRI revealed a large sphenoidal mucocele compressing on the superior and inferior orbital fissure. ESS proved to be a safe and effective treatment modality,
and when combined with early diagnosis and decompression, he was able to regain visual function.

**References**