CASE REPORT

Sphenoid mucocele with unusual panhypopituitarism

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SUMMARY

A 13-year-old boy presented with bilateral progressive proptosis, abduction deficit, optic atrophy and features suggestive of hypopituitarism secondary to a sphenoid sinus mucocele. Drainage of the mucocele along with hormone replacement therapy resulted in improvement in visual acuity and abduction.

BACKGROUND

Sphenoidal mucocoeles encompass a spectrum of manifestations, due to their close proximity to the structures enclosed by the orbital apex. The usual presentation includes irreversible diminution of visual acuity secondary to optic nerve compression and cranial nerve palsies. To the best of our knowledge, panhypopituitarism has not been documented with sellar-region mucocoeles. We present a case of a 13-year-old boy with bilateral progressive proptosis, abduction deficit, optic atrophy and systemic features of hypopituitarism, who was found to have a giant sphenoidal mucocele on radiological

evaluation. This case highlights the importance of maintaining a high level of suspicion about the possibility of a space occupying lesion and its systemic associations, especially when dealing with children. Moreover, prompt diagnosis and treatment can help in avoiding permanent visual loss.

CASE PRESENTATION

A 13-year-old boy presented with gradual progressive painless diminution of vision along with asymmetrical proptosis of both eyes (BE) for the past 3 years. There was a history of head injury after a fall from a moving vehicle 5 years prior, with no associated neurological or systemic symptoms. As per the history, no change in the proptosis was observed on forward bending, coughing or sneezing. On systemic examination, the patient appeared small for his age (height=112 cm) and had a deviated nasal septum due to a mass filling the right side of the nasal cavity. On ocular examination, the best-corrected visual acuity (BCVA) of the right eye (RE) and left eye (LE) was light perception with an accurate projection of rays in all quadrants and 1/60, respectively. Convergent deviation was noted in the RE, along with abduction deficit (−2) in BE. Furthermore asymmetrical proptosis was noted with values of 28 mm and 24 mm on Hertel's exophthalmometry in the RE and LE, respectively (figure 1). A relative afferent pupillary defect was noticed in the RE and fundus evaluation of BE revealed generalised disc pallor, suggestive of optic atrophy.

INVESTIGATIONS

Blood investigations reported values of haemoglobin, red blood cell count, cortisol, follicle stimulating hormone, luteinising hormone, testosterone

Figure 1 Clinical picture showing asymmetrical bilateral proptosis, widely placed orbits and abduction deficit in both eyes.

Figure 2 (A and B) CT scan and MRI, revealing large sphenoid sinus measuring 8.8×9.2×7.0 cm, extending craniocaudally from the region of the sella, sphenoid, ethmoid sinus, basifrontal anterior cranial fossa, nasopharynx, bilateral infratemporal fossa and nasal cavity.

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Figure 3 MRI scan showing marked separation of both bony orbits and narrowing of bilateral orbital apices (indicated by black arrow). The flow voids of the intracavernous part of both internal carotid arteries were widely separated (indicated by white arrow), in proximity to the expected pituitary gland location.

and adrenocorticotrophic hormone to be 6.6 g/dL, 2.8 million/ μ L, 5.5 μ g/dL, 0.230, 0.795, 0.025 mg/mL and 5.4 pg/mL, respectively. All these values were below normal range. The CT scan head and paranasal sinuses revealed a large hypodense peripherally enhancing lesion measuring 8.8 $\times$ 9.2 $\times$ 7.0 cm, which was seen extending craniocaudally along the regions of sella, sphenoid, ethmoid sinus, basifrontal anterior cranial fossa, nasopharynx, bilateral infratemporal fossa and nasal cavity (figure 2A). The orbits and nasal cavity were displaced anteriorly and nasally, along with shallowing of the orbits. Brain MRI was suggestive of a large well-defined T1-isointense and T2-hyperintense lesion filling and expanding the sphenoid sinus (figure 2B). Posterior pituitary bright spot and stalk could not be localised on MRI. The giant mass was seen completely obscuring the pituitary fossa. As a result, marked mass effect on

optic canals led to narrowing of the orbital apices and compression of bilateral optic nerves, which were presumably splayed over the mucocoele (figure 3).

DIFFERENTIAL DIAGNOSIS

The differentials of inflammatory orbital disorders, vascular malformations such as varices, carotid cavernous fistula, cystic lesions and tumours were considered.¹

TREATMENT

A diagnosis of sphenoid mucocoele with bilateral proptosis, abduction deficit, optic atrophy and panhypopituitarism was made. The patient was referred to the department of otorhinolaryngology for endoscopic drainage of the mucocoele along with administration of hormone replacement therapy (hydrocortisone and thyroxine). Complete evacuation of the mucocoele was noted on CT scans, postendonasal drainage (figure 4).

OUTCOME AND FOLLOW-UP

Visual acuity improved to 3/60 in the LE and abduction deficit resolved with fully regained ocular movements in BE at 4 weeks follow-up.

DISCUSSION

Paranasal sinus mucocoeles are epithelium-lined cystic masses.² These mucocoeles are seen more commonly among men, especially in the third or fourth decades.³ The incidence of sphenoid sinus mucocoele is reported to be 1–2%.⁴ Possible aetiological factors responsible for the formation of mucocoeles are trauma, tumour, inflammation, cranial dysplasias, chronic sinusitis and systemic disease.⁵ In our patient, there was a preceding history of significant head trauma that had occurred 2 years before the start of his ocular manifestations, thus the mucocoele may be attributed to trauma. Usually sphenoidal mucocoeles present with a variety of manifestations due to close vicinity to the orbital apex. Patients may present with chronic, severe headaches, supraorbital or retrobulbar in location, which get aggravated in

Figure 4 CT scan pictures showing complete evacuation of the mucocoele, postendoscopic drainage.

the supine position.⁶ Optic nerve involvement and cranial nerve palsies due to the compression effect over the orbital apex are common manifestations of sphenoidal mucocoeles, which may further lead to irreversible blindness.⁷ Visual field defects and endocrine dysfunctions have also been reported.⁸

Symptomatic relief in patients with sphenoid mucocoeles requires mandatory drainage of the mucocoele. The endoscopic approach is considered the 'gold standard', as it helps in better visualisation of the sphenoid sinus along with better restoration of respiratory function.⁹ Previous studies have shown poor visual outcomes, especially in cases with a sudden onset visual loss with BCVA of no light perception prior to drainage.¹⁰ Although panhypopituitarism in sphenoiditis has been reported earlier, sellar-region mucocoeles rarely manifest as hypopituitarism unless they directly originate from a paranasal sinus.¹¹

However, in our case, clinical manifestations such as visual disturbance, optic atrophy, abduction deficit as well as endocrine dysfunction were noted on examination. The parents had not noticed the unusual growth pattern of the child, nor had they seen the widely placed orbits and the child first presented to an ophthalmologist due to gradual decline in visual acuity. As it is not uncommon to miss subtle abnormal clinical features, it is essential for the parents as well as the ophthalmologist to not overlook any abnormal signs and symptoms. Ophthalmologists should assess minor symptoms with a high index of suspicion, especially while treating children. Identification and early intervention for abnormalities in such syndromic disorders with multiple system involvement would be helpful in preventing a permanent loss.

Patient parent's perspective

We had missed many signs and symptoms that indicated the multiple abnormalities in our child. We could have avoided irreversible damage if we had noticed them and brought our child to hospital earlier. Now, after surgery, we are hoping for improvement in the visual acuity and growth of our child.

To summarise, although ophthalmologists are primarily concerned with diseases of the eye, they should be more sensitive to identify any subtle abnormal systemic sign as well, thus aiding in prompt diagnosis and relevant management of the patient.

Learning points

- Maintain a high index of suspicion to identify unusual presentations.
- While evaluating the paediatric population, systemic examination including anthropometry should be mandatory.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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