Sphenoid mucocoele – an unusual cause for headaches in a teenage boy

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Isolated sphenoid sinus disease in childhood is uncommon and sphenoid mucocoeles (histologically benign, epithelium-lined, mucus-containing sacs) are rare. They are thought to arise as a result of obstruction of the sinus ostium due to previous surgery, inflammation, trauma or irradiation, but may be idiopathic. We present a case of a sphenoid sinus mucocoele in a young boy and highlight the fact that headache may be the only symptom, therefore diagnosis may require a combination of imaging and exploratory surgery.

CASE REPORT

A 15-year-old boy presented with retro-orbital headaches for 5 years, right peri-orbital swelling for 3 years and diplopia for several months. He had no symptoms of chronic sinusitis, allergic rhinitis or epistaxis. He had no history of cranial trauma or surgery and reported normal sensation of smell and vision. He presented to our otorhinolaryngology clinic in 2013, but was lost to follow-up until 2016.

On examination at his presentation in 2016, he was slight of build (like his mother) weighing 29.5 kg, which is below the 3rd centile for weight-for-age. He had right proptosis with infratemporal soft subdermal swelling (Figs 1A and 1B), normal visual acuity and no relative afferent pupillary defect. He experienced diplopia upon right lateral gaze, with reduced abduction and supraduction. There was reduced sensation in the distribution of the ophthalmic (V1) and maxillary (V2) branches of the trigeminal nerve. On nasal endoscopy, a pale, soft, submucosal mass was visible in the nasal cavity.

Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a sphenoid-based locally expansile cystic lesion (Figs 2 and 3). Extensive bony erosion and remodeling was noted. Extension intra-cranially with compression of the dura and cavernous sinus without invasion was noted. The lesion involved the orbital apex and extended into the pterygopalatine fossa and then through the pterygomaxillary fissure into the infra-temporal space where it was noted subdermally at the level of the zygoma.

Discussion

Sphenoid sinuses are small recesses in the postero-superior nasal cavity and remain intra-nasal until 6 - 7 years of age. Secondary pneumatisation occurs and a true cavity is formed by 8 - 10 years of age, with maturation by 12 - 14 years of age.[1]

Paranasal sinus mucocoeles are very rare in children and sphenoid mucocoeles make up only 1 - 2% of all paranasal sinus mucocoeles in all age groups.[2] To date, only 13 cases of paediatric sphenoid mucocoeles have been documented from 9 articles in the English literature.

Mucocoeles are histologically benign, epithelium-lined, mucus-containing sacs that affect the paranasal sinuses. They are thought to arise from obstruction of the sinus ostium due to previous surgery, inflammation, trauma or irradiation. Other potential aetiologies include cystic dilatation of glandular structures and cystic development from embryonic epithelial residues.[2] Increased levels of fibroblast
Presenting signs and symptoms are subtle until late in the disease process. In the literature, the most frequently reported manifestations are headache (89%), decreased visual acuity (57%), oculomotor palsies (56%) and exophthalmos (25%).[4,5]

In this case, the dysfunction of the ophthalmic (V1) and maxillary (V2) divisions of the fifth cranial nerve (trigeminal) is unusual and would make invasive lesions more likely. However, these cranial nerve dysfunctions due to a sphenoid mucocoele have previously been reported by Yong et al.[6] V1 may be compressed or stretched at the cavernous sinus or the orbital apex where it passes through the superior orbital fissure to enter the orbit. Similarly, V2 may be involved at the cavernous sinus, pterygopalatine fossa (which houses the pterygopalatine ganglion), the pterygomaxillary fissure en route to the infratemporal fossa, or in the roof of the maxillary sinus (infra-orbital nerve). This presentation of a mucocoele with isolated lateral facial swelling due to the soft tissue extension through the abovementioned spaces is the first such report to the best of the authors’ knowledge.

Mucocoeles may have variable densities on CT and signal intensities on MRI depending on their protein content, inspissation and presence of infection. MRI is best to delineate extent and internal characteristics of the lesion, whereas CT demonstrates bony involvement better.[3,5] The sinus walls may be thinned or completely dehiscent with extensive advancement into surrounding structures as in this case.

Although isolated sphenoid sinus disease in childhood is rare, the differential is broad and includes inflammatory conditions (e.g. sphenoiditis, polyposis, fungal sphenoiditis, hydatid cyst); congenital conditions (e.g. dermoid cyst, epidermoid cyst, meningocoele, meningoecephalocoele, Rathke’s cleft cyst, cranioopharyngioma); benign lesions (e.g. mucocoele, nasopharyngeal angiofibroma, osteoma, lymphangioma, haemangioma, pituitary tumours, internal carotid artery aneurysm, fibrodyplasia) and malignant sphenoid lesions (although they have not yet been reported in this age group, but considerations would be lymphoma, rhabdomyosarcoma, Ewing’s sarcoma).

Differentiating a mucocoele from these may be difficult with CT and MRI, as overlap in imaging characteristics is common. In this case, a meningocoele and meningomyelocoele were excluded after imaging showed no communication with intracranial spaces. The differential included a hydatid cyst (which was excluded on serology) and lymphangioma which could not be excluded. It may only be after surgery that a definitive diagnosis can be made. An external surgical approach was used in the past but an endoscopic transnasal route, with decompression of disease, has recently become the standard of care. This approach has a lower rate of complications and is effective regardless of size and pre-operative complications.[4]

**Conclusion**

Childhood sphenoid mucocoele is rare and the presenting symptoms may be vague. A headache associated with other neurological symptoms or signs warrants specialist review with endoscopy and/or further imaging. Combination CT and MRI is appropriate, but is not diagnostic in all cases. Even extensive or complicated disease is managed both safely and effectively with a diligent endoscopic transnasal approach.

**Learning points**

Isolated sphenoid sinus disease is very rare in children.

- A deep-seated, retro-orbital headache in a child is suggestive of sphenoid sinus disease and warrants further investigation, especially if associated with other neurological symptoms or signs.
CASE REPORT

• Cranial neuropathies must be excluded: optic > abducens > trigeminal > oculomotor > trochlear, as these may be indicative of invasive disease, and would warrant urgent referral.
• Imaging is helpful but may not be diagnostic.
• Endoscopic drainage is the treatment of choice for sinonasal mucocoeles and carries low morbidity.

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Fig. 4. Postoperative axial computed tomography (CT) indicating the lateral extent of the mucocoele (Lat), which is now aerated space. The sphenoid sinus septum (SSS) is pushed across to the left side and the eroded clivus (C). (B) Postoperative coronal CT with dehiscent middle cranial fossa (MCF) floor and zygoma (Z). (C) Postoperative sagittal CT. Note the clivus (Cl) erosion. (TNA = transnasal approach (surgical route of access); A = anterior; R = right; F = frontal sinus; ST = sella turcica; S = superior.)

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