Isolated Sphenoiditis Presenting with Abducens Nerve Palsy

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Abstract

Paranasal sinus infection is common in paediatric patients; however, isolated infection of the sphenoid sinus is rare and can result in life-threatening complications. Diagnosing Sphenoiditis on the basis of history and examination can be troublesome as the clinical picture is often non-specific and patients seldom complain of nasal symptoms. We report a case of sphenoid sinusitis in a 12-year-old boy who presented with unilateral VIth nerve palsy and visual disturbance. This case highlights the diagnostic challenge of sphenoid sinusitis and the importance of early collaboration between paediatric, ophthalmology and ENT specialists to achieve the best outcomes.

INTRODUCTION

Infection of the sphenoid sinus in isolation is rare and can be a challenge to diagnose as symptoms are often vague and non-specific. In addition, the sphenoid sinus frequently appears normal with endoscopic examination in the presence of infection [1,2]. Therefore, sphenoiditis is often misdiagnosed, which can lead to devastating complications if left untreated. We report an unusual case of isolated sphenoiditis in a 12-year-old boy who presented with right-sided VIth nerve palsy and visual loss.

CASE REPORT

A 12-year-old male patient was admitted to Bristol Royal Infirmary, UK with a 3-month history of progressive visual disturbance and diplopia. There was no associated fever, nausea, vomiting, photophobia, or phonophobia. He had an MRI scan 6 weeks prior to his admission at another local hospital, which was reported as normal. He had two episodes of headache in the week prior to admission but was otherwise generally well and attending school as normal. On examination, incomplete right abducens nerve palsy was noted. CT scan confirmed right-sided sphenoid infection and showed an abnormality in the region of the right petrous apex and clivus with a small bony irregularity suggestive of an inflammatory process (Figure 1). Repeat MRI of the brain, orbits and sinuses showed opacification of the right sphenoid sinus as well as right cavernous sinus and inferior petrosal sinus thrombophlebitis (Figure 2, 3).

The patient was placed on intra-venous Metranidazole and Cefuroxime. Vistamethasone and Xylometazoline hydrochloride nasal drops were also prescribed. He underwent elective right-sided sphenoidotomy and a small amount of pus was collected. Culture of the pus showed no bacterial growth. Subcutaneous Enoxaparin was administered during the time he was an inpatient (6 days). Anticoagulation was not continued after discharge as his symptoms had significantly improved by the fourth day of admission and treatment of the underlying infection was deemed sufficient over the risks of long-term anticoagulation in a child.

2 months after surgery, his visual acuity had fully recovered however a subtle sixth nerve palsy and diplopia at middle and far distances persists. He remains well at school and wears an eye patch as and when required. Repeat MRI scan showed resolution of cavernous and inferior petrosal sinus thrombophlebitis.

Figure 1 Axial CT scan showing opacification of the right sphenoid sinus.
DISCUSSION

In recent years, the incidence of sphenoid sinusitis has been rising, which is probably as a result of increasing use of CT and MRI scans. Sphenoid sinusitis accounts for only 2.7% of all sinus infections and isolated sphenoiditis is very rare with an approximate incidence of 0.4% in children/adolescents [3,4]. The sphenoid sinus is lined with ciliated pseudostratified epithelium with fewer mucous secreting cells as compared to other paranasal sinuses [5]. This causes less drainage problems and may explain the relative rarity of isolated infection.

The dangers associated with sphenoid infection can be attributed to its anatomical position as it lies in close proximity to many vital structures including: the pituitary gland, optic nerve, internal carotid arteries, cavernous sinuses, sphenopalatine nerve, sphenopalatine ganglion and cranial nerves III-VI (Figure 4) [6-8]. Cadaveric studies carried out by Fuji et al. [9] demonstrate that in 78% of specimens the bony sphenoid wall was less than 0.5mm in thickness. 4% of specimens had only the optic sheath and sinus mucosa separating the nerves from the sinus. The anatomical location and relatively thin walls of the sphenoid sinus contribute to an increased risk of complications arising as a result of intracranial spread of infection.

The sixth cranial nerve lies medial to the cavernous sinus and is most commonly affected. It takes a long intracranial course as it exits the brainstem from the inferior pontine sulcus and passes along the bony clivus through to Dorello’s canal. It then goes through the cavernous sinus before entering the orbit via the superior orbital fissure. The abducens nerve is particularly vulnerable to damage in the region of the petrous apex as a result of progressive thromboophlebitis affecting the inferior petrosal sinus [10,11].

Symptoms of paranasal sinusitis including nasal obstruction, discharge, postnasal drip, hyposomia and facial pain are rarely experienced by those with sphenoiditis. Review of the literature suggests that headache is the most common symptom (85-98%) [6,12]. A retrospective study of 61 patients carried out by Gilonyet al. [12] report headache location in descending order of frequency: frontal, vertex, occipital, posterior parietal and retro orbital. Visual blurring or loss of visual acuity constitutes the second most common symptom [13]. Our patient presented with diplopia and visual disturbances. There were no other sinus related complaints apart from a vague history of two isolated episodes of headache, two weeks prior to his admission. Previously reported cases of sphenoiditis with sixth nerve palsy report chronic headache of varying severity which accompany visual changes [14-16]. In this case, the presentation is particularly unusual as the eye changes occurred in the absence of other significant symptomology.

A high index of suspicion is required when suspecting sphenoiditis, particularly in children and adolescents with persistent headache that does not respond well to conventional symptomatic treatment. The presence of neurological findings suggests an intracranial complication, which should be investigated immediately. Flexible nasendoscopy can identify sphenoid infection if pus is present in the middle meatus or on the anterior surface. Oedematous or polypoidal changes in the sphenoethmoidal recess also imply sinus infection [17]. However the sensitivity of detecting sphenoid pathology via endoscopy is only 45% according to Nour et al.[1] High resolution CT scanning is the diagnostic tool of choice as it reveals sinus opacifications and delineates the bony anatomy [2]. On the other hand, MRI is superior when intracranial symptoms are present as it reveals soft tissue involvement [7].

Predisposing factors for sphenoiditis include Structural abnormalities (small sphenoid ostia, septal deviation, hypoplastic sinuses [4]), Allergic rhinitis, nasal/sinus polyps, Infective causes (URTI, direct spread from ethmoid sinusitis, swimming or diving with forceful water entry into the nose). Other causes include neoplastic disease, impaired mucociliary clearance (Kartanger’s syndrome and cystic fibrosis), immunosuppression, craniofacial radiotherapy and cocaine use [18,19]. However, none of these factors could be found in our patient’s history.

The pathogens most commonly involved in sphenoidal infection are *Staphylococcus aureus* followed by various *Streptococcal* species [20]. *Chlamydia, Klebilla, Haemophilus influenzae* and community acquired MRSA are examples of gram-negative and anaerobic organisms that are infrequently cultured [21]. Fungi, especially *Aspergillus* must be considered in all cases, particularly if the patient is immune compromised [5].
Differential diagnoses include primary central nervous system neoplasms that may compress the sixth nerve anywhere along its path. Any cause of elevated intracranial pressures may cause traction of the nerve at the skull base resulting in abducens nerve palsy. Trauma to the head, particularly orbital fractures can damage the neuronal structures. Other pathology affecting the sixth cranial nerve includes: Multiple sclerosis, encephalitis, meningitis and congenital causes [22].

The prognosis of sphenoiditis is highly variable; however prompt treatment with broad spectrum antimicrobial therapy, in uncomplicated cases, has a very good outcome. Topical decongestants and saline irrigation are also advised to help promote sinus drainage. The exact antibiotic regime will depend on local policy and the hospital microbiologist should be consulted. Complicated infection requires ENT specialist referral and subsequent sphenoidectomy for surgical drainage of the sinus. Haematology involvement is necessary in the case of cavernous sinus thrombosis for the addition of anticoagulant and anti-platelet aggregation therapy.

CONCLUSION

Isolated sphenoid infection is rare and can lead to life threatening complications if left untreated. A high index of suspicion is required when assessing children or adolescents with headache that does not respond well to conventional symptomatic treatment. Early collaboration between paediatric medical and ENT surgical teams, along with haematologists and ophthalmologists in complicated cases is necessary to achieve the best outcomes.

REFERENCES


