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Series of isolated sphenoid disease: Often neglected but perilous

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Abstract

Isolated sphenoid sinus disease (ISSD) is an uncommon entity and may present with non-specific symptoms. As sphenoid sinus is located adjacent to vital structures, a diseased sphenoid sinus can potentially lead to devastating complications such as cranial neuropathies, cavernous sinus thrombosis and intracranial abscess. Herein, we report a case series of three patients who presented with different localization of headache as their main symptom. Endoscopic nasal assessment showed abnormal nasal cavity findings with mucosal thickening and soft tissue mass occupying the sphenoid sinus revealed by computed tomography (CT) scan. All the sphenoid sinuses were drained via endoscopic approach. Post operatively, antibiotics or anti-fungal were given, however, these three cases displayed the possible variation in severity of this disease. Case 1 had an uncomplicated inflammatory disease of the sphenoid sinus, which is of bacterial origin, while the other two cases had fungal infection. Case 2 achieved disease stability with anti-fungal treatment, but Case 3 had a serious complication after treatment with permanent vision loss. A patient who presents with atypical headache, not responsive to analgesia should be investigated promptly with nasal endoscopic examination and radiological imaging for isolated sphenoid sinus disease to avoid devastating complications.

Keywords

Sphenoid sinus, sinusitis, headache, endoscopic sinus surgery

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Introduction

Isolated sphenoid sinus disease (ISSD) is a relatively rare condition. It is defined as disease localized only to the sphenoid sinus owing to several conditions, including inflammation, fungal disease and tumors.[1] These patients are usually not referred to the otorhinolaryngologist (ORL) early as they commonly present with non-specific headache.[1,2] The location of the sphenoid sinus at the base of skull increases the difficulty of diagnosis as it is not easily visualized on conventional radiograph. However, advances in imaging modalities in the current era has potentially led to an increase in diagnosis of ISSD.[1] There are many vital structures in the vicinity of the sphenoid sinus. Delay in diagnosis puts vital neurovascular structures at risk, hence prompt detection and early treatment is crucial to avoid catastrophic complications.[3] Here, we present three cases of ISSD who presented with headache but of different disease severity.

Case series

Case 1: A 41-year-old Chinese gentleman with no comorbidities presented with 4 months history of throbbing headache in the occipital and retro-orbital region, worsened when bending forward. He also complained of posterior nasal drip but did not have any other significant nasal symptoms. Nasoendoscopy showed mucoid discharge from the left sphenoid sinus suggestive of sphenoid sinusitis. Computer tomography (CT) scan confirmed the diagnosis with evidence of mucosal thickening occupying the left sphenoid sinus (Figure 1). He underwent a left endoscopic transnasal sphenoidotomy. Intraoperative culture was positive for *Citrobacter* sp. Post operatively, he was treated with 1 week of intravenous (IV) combination of amoxicillin and clavulanic acid after which his symptoms completely resolved. He remained well during his follow up over a period of 1 year.

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Case 2: A 61 -year-old Chinese gentleman with underlying hypertension and transient ischemic attack, came with complains of 2 months of right-sided headache at the frontal area with no other nasal or visual symptoms. Nasoendoscopy showed polypoidal soft tissue mass at the right sphenoethmoidal region. CT scan showed a contrast-enhancing lesion in the right sphenoid recess involving the right cavernous sinus through a bony dehiscent at the superior sphenoid wall (Figure 2). Magnetic resonance imaging (MRI) brain scan showed no evidence of intracranial extension. Biopsy taken from the nasal mass reported as fungal infection of *Mucor* sp. He was then treated aggressively for invasive fungal sinusitis. He underwent right sphenoidotomy with frequent nasal toileting twice per week under endoscopic guidance in the clinic and concurrent IV amphotericin B for 2 weeks. He was then put on oral posaconazole for a total duration of 1 year. He no longer complains of headache by the time of 1 month of anti-fungal therapy. Follow up CT and MRI scans 12 months post-surgery showed no evidence of fungal infection. Clinical follow-up with nasoendoscopy showed clear right sphenoid sinus with no evidence of recurrence.

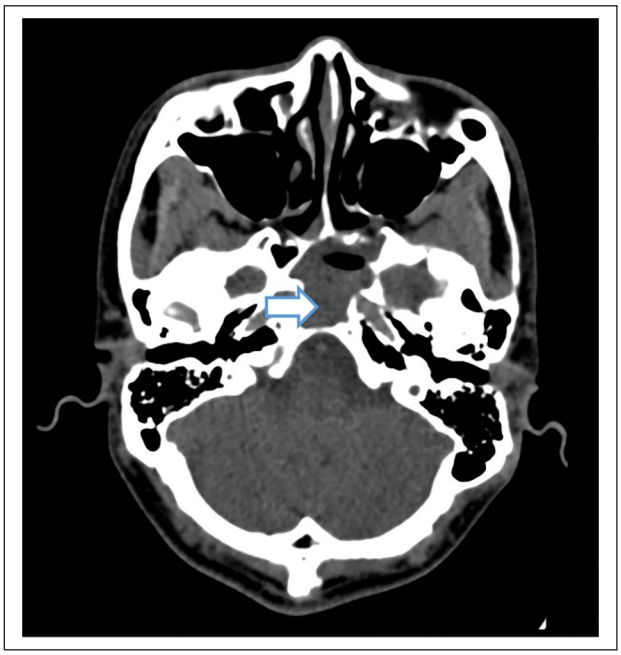

Figure 1. Axial slice CT paranasal sinus at the level of sphenoid sinus with white arrow showing mucosal thickening within the dominant left sphenoid sinus.

Case 3: A 56-year-old Malay lady with underlying diabetes mellitus, hypertension and ischemic heart disease sought treatment for intermittent left-sided pulsatile headache for 1 month. She was initially treated as left temporal arteritis by medical team. She visited the emergency department when her symptoms deteriorated with acute left eye pain, diplopia and blurring of vision. She also had intermittent left-sided yellowish rhinorrhoea. Clinically, she had a left eye proptosis with complete ophthalmoplegia. Nasoendoscopy showed yellowish mucoid discharge from sphenoethmoidal recess. CT scan showed mucosal thickening of the left sphenoid sinus with hyperdensities seen within (Figure 3). Due to the deteriorating vision, she underwent an emergency left endoscopic sphenoidotomy, during which blackish fungal debris were seen occupying the left sphenoid sinus. Intraoperative yielded fungal ball cultured positive for *Mucor, Rhizopus* and *Aspergillus* sp. She was co-managed by the infectious diseases team and put on a lifelong anti-fungal treatment because of remnant soft tissue lesion in the sphenoid sinus as well as persistent left internal carotid artery thrombosis seen in her post-operative MRI scan. During follow up visits, she

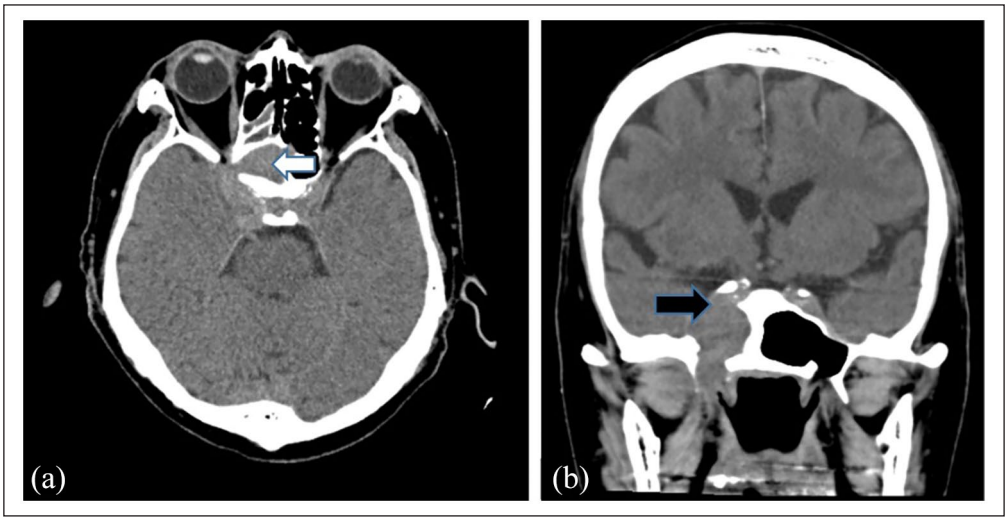

Figure 2. (a) Axial slice CT paranasal sinus at the level of sphenoid sinus with white arrow showing contrast-enhanced lesion within the right sphenoid sinus with involvement of cavernous sinus. (b) Note the bony dehiscence on coronal slice at the superior sphenoid wall, pointed out with black arrow.

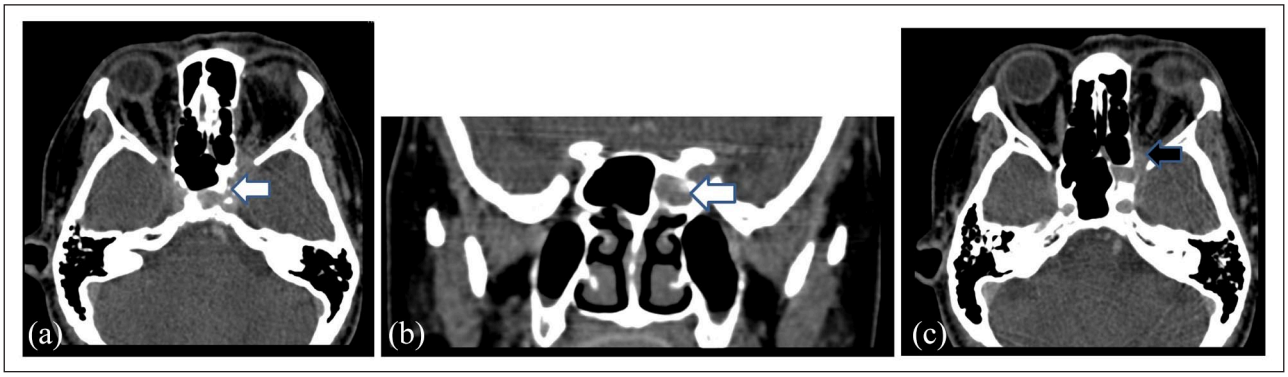

Figure 3. (a, b) Axial and coronal slice CT paranasal sinus at the level of sphenoid sinus with white arrow showing hyperdensities within the mucosal thickening in the left sphenoid sinus. (c) The axial slice clearly demonstrated the pathological involvement of left orbital apex (obliterated fat, black arrow) as compared to the normal right side.

no longer complained of headache or any nasal symptoms; however, her left eye vision loss remained permanent. Her CT and MRI scans 2 years post-surgery showed minimal residual soft tissue lesion which remained stable and unchanged.

Discussion

Headache is a common presenting symptom and has a high impact on society due to its disabling nature, leading to reduced quality of life and increased rate of absenteeism from work or school. A questionnaire-based study by Yoon MS et al. in Germany reported the prevalence of headache in up to 58% of subjects.[4] This is similarly seen worldwide, Stovner et al.[5] included 107 studies across all continents reported that 47% of the population globally is actively suffering from headache. Despite the high prevalence rate worldwide, headache disorders remain under-recognized and inadequately treated.

Headache is present in 65%–98% of patients with ISSD.[6,7] International Headache Society have classified sinusitis as one of the causes of secondary headaches. This disease is a rare entity and can present with headache as the sole symptom. Contrary to usual rhinosinusitis cases, only 20% of patients with ISSD complain of associated nasal or ophthalmologic symptoms.[6] In our case series, all 3 patients presented with different localization of headache including frontal, occipital, retro-orbital headache and pulsatile headache. This is unsurprising as literature described variable distribution of location of headache in ISSD.[8] ISSD can sometimes be mistaken for migraine or tension-type headache due to its various localization of headache. This poses a diagnostic challenge for the medical personnel, as demonstrated in our Case 3 whereby the patient was initially treated provisionally as left temporal arteritis by general medical team with analgesics for about a month, before referral to the ORL team.

Due to its anatomic location, the sphenoid sinus is often overlooked.[7] A high degree of suspicion is needed, and an office endoscopic nasal examination should be performed. Celenk reported positive nasoendoscope findings including nasal polyps, nasal secretions, mucosal edema and hemorrhagic crusts in 70% of patient with ISSD.[8] However, Socher et al.[9] and Nour et al.[10] failed to detect any pathological abnormalities by endoscope in more than half of the cases. Therefore, normal findings during nasoendoscopy do not exclude the presence of ISSD. This suggest that imaging tests are necessary to diagnose sphenoid diseases.

If headache of sphenoidal origin is suspected, a CT or MRI imaging should be done to accurately and timely detect ISSD. A variety of pathologies can cause ISSD including inflammation, benign or malignant neoplasm, osteofibrotic disease and vascular lesions. The choice of imaging will depend on the suspected pathology. CT has a 95% sensitivity in diagnosing inflammatory lesions and 100% sensitivity in diagnosing fungal sinusitis due to the presence of characteristic intrasinus mottling.[11] If sphenoid sinus malignancy is suspected, both CT and MRI play an equally important role in assessment as CT is more superior in defining bony erosions while MRI allows better mapping of the lesion.[11]

A retrospective analysis by Ashida et al.[1] discovered that 78% of patients with radiographic evidence of sphenoid sinus opacification were not referred to an otorhinolaryngologist for a thorough examination. As this study was conducted retrospectively based on radiology reports, the findings may have been incidental in an asymptomatic patient. However, due to the proximity of the sphenoid sinus to various vital structures, doctors should not underestimate the risk of sphenoid sinus disease and to leave a detected problem untreated.

All three cases described in this article presented with headache with one case having visual loss. This is coherent with a review by Fooanant et al.[12] where 63.9% of patients had headache and visual loss came in second. The trend differs in pediatric population, as they tend to present with headache and fever as the primary symptom.[13] We had one case of bacterial and two cases of fungal infection in our series. Inflammatory etiology contributes 75% of ISSD,

including bacteria, fungus, mucocele, retention cyst and polyp.[12] Despite inflammatory conditions being the most common, caution is needed when evaluating the sphenoid sinus in both symptomatic and asymptomatic patients. Knisely et al.[14] reported an incidence of 10.9% malignancy among patients with isolated sphenoid sinus opacification. Therefore, with the guidance from endoscopic assessment and high sensitivity imaging evaluation, a biopsy of the sphenoid sinus lesion should be done when indicated to achieve a pathological diagnosis and enable targeted treatment.

In the current era, endonasal endoscopic sphenoidotomy has gained favor over open approaches. Endonasal endoscopic sphenoidotomy showed satisfying results with a high cure rate of 92.2% [6] and significant improvement in comparison of pre and post-operative mean VAS score for headache.[8] Fadda et al.[15] also reported great survival and complete remission rate with concurrent antibiotics therapy and early surgical debridement. All three of our patients underwent endoscopic sphenoidotomy via transethmoidal approach, followed by appropriate antibiotics. All three patients showed positive results and resolved headache.

Conclusion

The diagnosis of ISSD requires high index of clinical suspicion as presenting symptoms are usually nonspecific. Headache is the commonest presenting symptom albeit variable in location. Clinicians should suspect ISSD in patients with vague intractable headache. Advances in imaging have aided and improved the detection of ISSD. All doctors should have sufficient knowledge of sphenoid sinus lesions and not underestimate its risk. If a sphenoidal sinus lesion is found incidentally on imaging, a closer examination by an otolaryngologist is recommended.

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Ethical approval

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Informed consent

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