International Journal of Otorhinolaryngology and Head and Neck Surgery
Fayad G et al. Int J Otorhinolaryngol Head Neck Surg. 2024 Oct;10(5):541-544
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20242286

Case Report

Staphylococcus epidermidis isolated sphenoiditis: a rare cause of recurrent orbital apex syndrome

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Received: 28 July 2024
Accepted: 12 August 2024

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ABSTRACT

Isolated sphenoid sinusitis (ISS) is a rare condition that can lead to severe complications, with orbital apex syndrome (OAS) being an exceptionally rare manifestation characterized by visual loss and multiple cranial neuropathies. We report a case of a 41-year-old female presenting with recurrent OAS caused by a chronic *Staphylococcus epidermidis* infection of the sphenoid sinus. Despite extensive diagnostic workups and multiple surgical interventions, the patient experienced recurrent episodes of OAS, highlighting the challenges in diagnosis and treatment. This report emphasizes the pathogenic potential of *S. epidermidis*, typically considered a commensal organism, in chronic sinusitis and underscores the importance of a multidisciplinary approach in managing such complex cases.

Keywords: Isolated sphenoid sinusitis, Orbital apex syndrome, *Staphylococcus epidermidis*, Recurrent infection, Cranial neuropathies, Visual loss

INTRODUCTION

Isolated sphenoid sinusitis (ISS) is a rare condition that can lead to severe complications, with orbital apex syndrome (OAS) being an exceptionally rare manifestation characterized by visual loss and multiple cranial neuropathies.[1,2] We report a case of a 41-year-old female presenting with recurrent OAS caused by a chronic Staphylococcus epidermidis infection of the sphenoid sinus, highlighting the diagnostic and treatment challenges due to the recurrent nature of the disease and involvement of this typically commensal pathogen.

CASE REPORT

A 41-year-old female with no significant medical history presented to the emergency department with fever, acute unilateral left-sided severe fronto-temporal headache, left-eye vision loss and retro-ocular pain, diplopia, and left-sided facial numbness. The symptoms developed rapidly over several days. Physical examination revealed ptosis of the left upper eyelid, binocular diplopia, reduced visual acuity and complete ophthalmoplegia of the left eye, hypoesthesia in the distribution of the ophthalmic branch of the left trigeminal nerve (V1).

Initial imaging with an angiographic computed tomography (CT) scan of the head ruled out any cerebro-vascular anomalies (e.g., carotid cavernous aneurysm, carotid cavernous fistula, cavernous sinus thrombosis) and showed only an isolated left sphenoid sinusitis with thickening of the sphenoid sinus wall, suggestive of chronic inflammation (Figure 1).

Subsequent magnetic resonance imaging (MRI) demonstrated T1 contrast-enhanced hypersignal in the left sphenoid sinus with extension to the anterior region of the cavernous sinus and towards the orbital apex (Figure 2).

Notably, there was no evidence of cavernous sinus thrombosis, orbital cellulitis, or invasive fungal infection, confirming the diagnosis of isolated sphenoid sinusitis with orbital apex involvement.

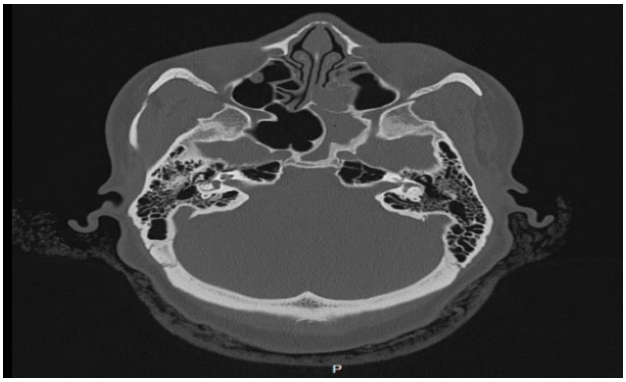

Figure 1: Pre-operative axial CT-scan showing acute left sphenoid sinusitis with bone thickening (hyperostosis), indicative of chronic sinusitis.

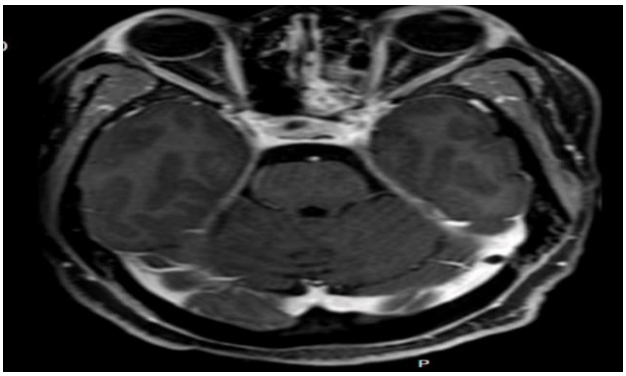

Figure 2: Pre-operative MRI T1-weighted image with contrast (Gd) showing a hypersignal left sphenoid sinusitis with extension to the anterior cavernous sinus and left orbital apex.

Initial management included empiric intravenous ceftriaxone 2 g daily and high-dose methylprednisolone for two days. A unilateral extended sphenoid sinus antrostomy as part of functional endoscopic sinus surgery (FESS) was performed. Initial microbiological cultures of sinus aspirates isolated Staphylococcus epidermidis, which was initially ruled-out as a commensal. Aerobic, fungal, and mycobacterial cultures were negative. The patient recovered well, her symptoms gradually disappeared, and her visual acuity was restored. She was closely monitored with regular debridement of the surgical site.

However, the patient experienced recurrent episodes of OSA characterized by fever, severe headaches, visual loss in the left eye, and left cranial nerve palsies (III, V1, VI). Nasofibroscopy showed restenosis of the left sphenoid ostium and a repeat CT-scan showed recurrent left sphenoid sinusitis. More aggressive surgical interventions were required, including bilateral extended sphenoid antrostomy (sphenoid "drill-out") and ultimately a radical sphenoidectomy with a naso-septal flap (Figure 3).

After each surgery, the patient received intravenous corticosteroids and antibiotics, primarily ceftriaxone.

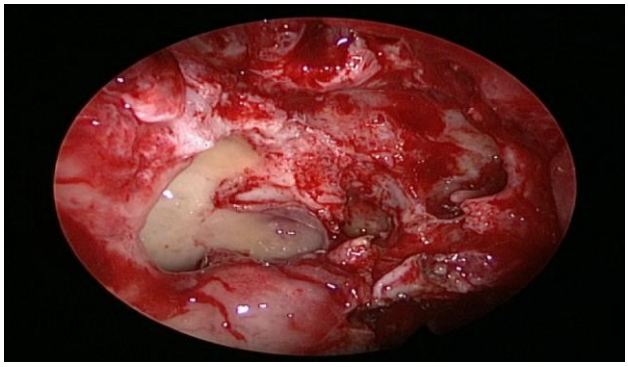

Figure 3: Intra-operative endoscopic view of the radical sphenoidectomy with wide opening of the sphenoid sinuses and a naso-septal flap on the sinus floor.

Histopathological analysis was suggestive of nonspecific chronic inflammation and ruled out granulomatosis, vasculitis, neoplastic processes, invasive aspergillosis or mucormycosis, and IgG4-related disease. An extensive diagnostic workup was conducted, including comprehensive blood tests for systemic conditions (e.g., tuberculosis, autoimmune markers, IgG4 and ACE enzyme levels), which were all negative. Serum laboratory studies ruled out immunosuppression and diabetes mellitus. Ophthalmological assessment confirmed retro-bulbar optic neuritis via visual evoked potential testing, which was treated again with high-dose corticosteroids. The patient unfortunately developed cortico-induced glaucoma, exacerbating her visual loss, and which was managed appropriately. Neurological evaluation, including brain MRI and lumbar puncture, excluded multiple sclerosis, meningitis, and other possible central neurological causes.

Despite the initial challenges and multiple surgical interventions, the patient achieved total recovery of all cranial nerve palsies and complete resolution of retro-bulbar optic neuritis. Follow-up imaging and nasal examination at 6 months post-operatively showed complete resolution of the sphenoid sinusitis (Figure 4).

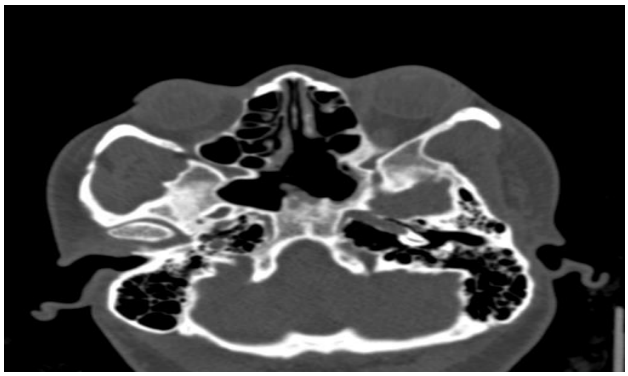

Figure 4: Post-operative axial CT-scan after radical sphenoidectomy showing resolution of the left sphenoid sinusitis with resolution of bone thickening.

DISCUSSION

ISS is a relatively rare condition, accounting for only 1-2% of all paranasal sinus diseases.[1] ISS typically presents with symptoms such as headache and retro-orbital pain (82%), and ocular symptoms (28%) including reduced visual acuity, ophthalmoplegia, and diplopia. Common risk factors for ISS include immunosuppression, diabetes, exposure to contaminated water, cocaine use, steroid therapy, and craniofacial abnormalities.[1] However, our patient had none of these risk factors. The most frequently isolated pathogens in ISS are *Staphylococcus aureus*, *Streptococcus* species, and *Aspergillus*.[3] Despite its rarity, the sphenoid sinus's anatomical location at the skull base, adjacent to critical structures, can lead to significant complications including meningitis, subdural abscess, cavernous sinus thrombosis, orbital infection, and, more rarely, ocular cranial nerve (CN) palsies (primarily CN VI followed by CN III).[4] Although cranial nerve palsy is typically indicative of a neoplastic process within the sphenoid sinus, it can also be associated with aggressive bacterial sinusitis, allergic fungal sinusitis, or invasive fungal infections.

OAS is another uncommon but serious condition characterized by multiple cranial neuropathies affecting CN II, III, IV, V1, and VI, at the level of the orbital apex. Patients with OAS typically present with ptosis, proptosis, visual loss, ophthalmoplegia, and hypoesthesia in the distribution of the ophthalmic branch of the trigeminal nerve.[2] While the etiologies of OAS include infectious, inflammatory, neoplastic, traumatic, and vascular processes, invasive fungal sinusitis and orbital cellulitis are the most commonly implicated causes. These conditions are associated with high morbidity and mortality rates, necessitating a high index of suspicion for patients presenting with OAS.

The association between bacterial sinusitis and OAS, particularly without concurrent orbital cellulitis, is extremely rare. Our literature review identified only twelve such cases, with only three specifically involving ISS.[5-14] The pathogens in these cases were often polymicrobial and included methicillin-sensitive *Staphylococcus aureus* (MSSA) (2 cases), *Pseudomonas aeruginosa* (4 cases), *Klebsiella Rhinoscleromatis* (1 case), and *Staphylococcus epidermidis* (1 case). Immunosuppression and diabetes mellitus were present in most patients (10/12), which probably played an important role in the rapid progression and severity of the disease.

In our reported case, repeated microbiological cultures consistently showed heavy growth of *Staphylococcus epidermidis*, and our extensive work-up ruled out any other possible pathogen or cause. Given the recurrent and temporal association between our patient's clinical, biological, and radiological presentation of ISS with OAS, and only after ruling out all other possible causes, we considered this to be a true *S. epidermidis* infection. Typically considered a low-virulence commensal, *S. epidermidis* can act as an opportunistic pathogen under certain conditions, such as in immunocompromised and diabetic individuals.[15] The pathogenicity of *S. epidermidis* in chronic sinusitis and its potential role in causing OAS can be attributed to its capability of forming biofilms.[16] These biofilms enhance the bacterium's ability to adhere to surfaces and resist antibiotic treatment, leading to chronic and difficult-to-eradicate infections. In our patient, the recurrent nature of the sphenoiditis and stenosis of the sphenoidotomy site suggest that biofilm formation may have played a significant role.

Management of OAS due to bacterial sinusitis aims to stop the extension of the infection intracranially and involves a combination of broad-coverage antibiotic therapy, early surgical drainage, and a high index of suspicion for invasive fungal infection. Our patient experienced recurrent restenosis and sinusitis and underwent multiple surgeries, including unilateral and bilateral extended sphenoid sinus antrostomies and a radical sphenoid-dectomy, ensuring drainage and removal of infected tissue. Between surgeries, the patient received intravenous corticosteroids and antibiotics (primarily ceftriaxone), nasal irrigation, and regular debridement of surgical site. Ophthalmological management is also a critical for the patients' care, as ISS can lead to visual impairment, with or without OAS, through contiguous spread of infection and inflammation to the optic nerve.[4] Notably, in almost all reported cases (11/12), patients suffered irreversible visual loss despite appropriate treatment. highlighting the high risk associated with OAS from isolated bacterial sinusitis even in the absence of orbital cellulitis. However, our case is the first to demonstrate complete visual acuity recovery, even after multiple disease recurrence and treatments. Monitoring for cortico-induced glaucoma, especially during the treatment of retro-bulbar optic neuritis, was vital to prevent further vision loss and manage the side effects of prolonged corticosteroid use.

CONCLUSION

Isolated bacterial sinusitis, specifically ISS, can lead to severe complications such as OAS, with high risk of permanent visual loss. *Staphylococcus epidermidis*, a nasal commensal, has the potential to act as an opportunistic pathogen in chronic sinusitis, potentially leading to ISS and ultimately OAS. This case highlights the potential for OAS to arise from bacterial sinusitis, emphasizing the need for thorough diagnostic evaluation, early recognition, aggressive treatment, and a multidisciplinary approach.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Fayad G, Cavelier G, Verhasselt M, Rodriguez A, Horoi M. *Staphylococcus epidermidis* isolated sphenoiditis: a rare cause of recurrent orbital apex syndrome. Int J Otorhinolaryngol Head Neck Surg 2024;10:541-4.