



A Rare Case of Fungal Rhinosinusitis Caused by *Scedosporium apiopermum*

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Abstract

Fungal rhinosinusitis (FRS) is inflammation of the paranasal sinus mucosa due to fungal infections, which can be invasive or non-invasive. The occurrence of a sphenoid mucocele with a fungal ball is rare. We report a case of sphenoid sinus mucocele with a fungal ball caused by *Scedosporium apiopermum* in a 32-year-old female who presented to the Emergency Department with persistent headache not relieved on medications. The radiological images showed a mucocele with clival osteomyelitis. Urgent endoscopic examination and debridement was undertaken which demonstrated a mucocele with fungal ball. Microbiological examination confirmed it to be *Scedosporium apiopermum*.

Keywords Invasive fungal sinusitis · Non-invasive fungal sinusitis · Mycetoma · Fungal ball · Sphenoid mucocele · Skull base osteomyelitis · *Scedosporium apiopermum* · *Aspergillus*

Introduction

Fungal rhinosinusitis (FRS) may be defined as inflammation of the paranasal sinus mucosa due to fungal infections. It is categorised as either non-invasive (fungi confined to the lumen of sinus) or invasive (fungi breach the epithelial lining to infiltrate the mucosa and underlying connective tissues, including vessels). The distinction is clinically significant: invasive FRS classically presents in immunocompromised patients and has a more aggressive clinical course than non-invasive FRS; early diagnosis and treatment are critical to improved clinical outcomes.

There are two forms of non-invasive fungal rhinosinusitis: mycetoma (fungal ball) and allergic fungal rhinosinusitis. The latter is defined as an intense, localized allergic/eosinophilic inflammatory disease that results in the accumulation of eosinophilic (allergic) mucin (a thick, tenacious eosinophilic secretion that contains fungal hyphae), intense eosinophilic inflammation, and characteristic radiographic findings. The maxillary sinus is the most commonly affected sinus.

Mucoceles are benign, expansile, cysts and may involve the paranasal sinuses and become secondarily infected. Bacterial infection is most common, resulting in the formation of a pyocele. By contrast, fungal infection of mucoceles is rare. *Aspergilli sp.* are the most common cause of fungal infection in the sinonasal tract, both for invasive and non-invasive cases [1].

We present a rare case of sphenoid sinus mucocele with mycetoma formed by *Scedosporium apiopermum*. Fungal disease has an unpredictable presentation which warrants careful assessment and radical treatment.

Case Report

A 32-year-old female presented to the Emergency Department (ED) in a tertiary care hospital in Birmingham with a one-month history of headache, dizziness and intermittent loss of consciousness. She did not have any comorbidities and clinical examination was unremarkable.

A computed tomography (CT) scan of the skull base showed expansile opacification of the left hemisphenoid cavity with focal bone erosions at multiple sites. There were also features of osteitis of the surrounding bone and bowing of the intersphenoid septum towards the left. Posteriorly, the clivus was eroded up to the dural surface on the

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left side of midline (Fig. 1). The radiological appearances were suggestive of a sphenoid sinus mucocoele; the working clinical diagnosis was of headache secondary to the expansile nature of the mucocoele. Initial management was with oral analgesia, however, her symptoms worsened and she represented to ED with more severe episodes of dizziness, which were now associated with diplopia and photophobia. A post-contrast T1 Magnetic Resonance Imaging (MRI) confirmed the previous CT findings: there was evidence of peripheral enhancement which was suggestive of mucocoele rather than neoplasia (Fig. 2a, b). Interestingly, however, her T2 imaging revealed a signal void within the mucocoele

(Fig. 2c). The Patient was referred for ophthalmological opinion, which noted that her pain was predominantly in the left periorbital region radiating to her forehead. She also had esotropia (medial deviation of the eye) and left lateral rectus weakness, probably due to her clival erosion with pressure effect on the abducens nerve.

An urgent endoscopic examination and debridement was undertaken which showed polypoidal mucosa over the left superior sphenoidal recess. A wide endoscopic sphenoidectomy was performed which revealed a large cauliflower-shaped mycetoma (Fig. 3) with an additional mucocoele in the left sphenoid cavity. The mycetoma was removed and

Fig. 1 (a) Computed Tomography Scans: expansile opacification of the left hemisphenoid cavity with focal bone erosions at multiple sites. Arrows mark sites of bone erosions. (b) Posteriorly, the clivus is partially eroded up to the dural surface on the left side of midline. Parietal dura has a smooth bulge

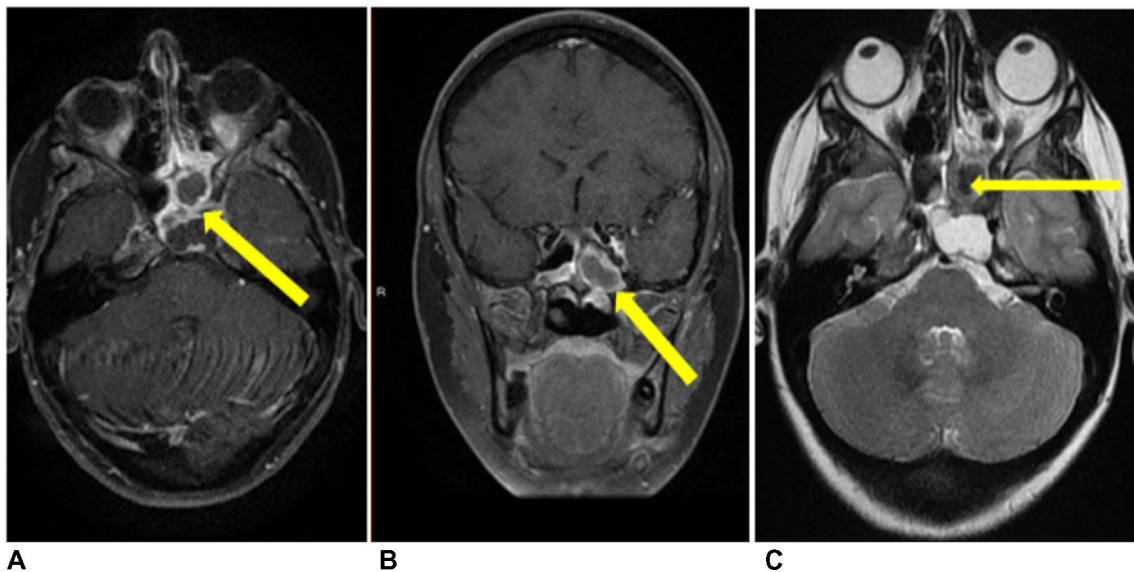
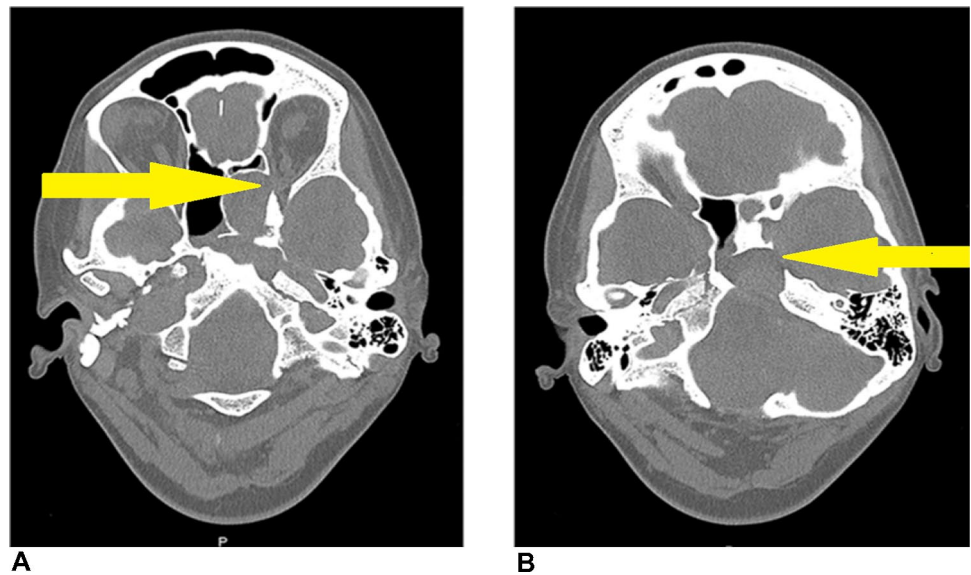
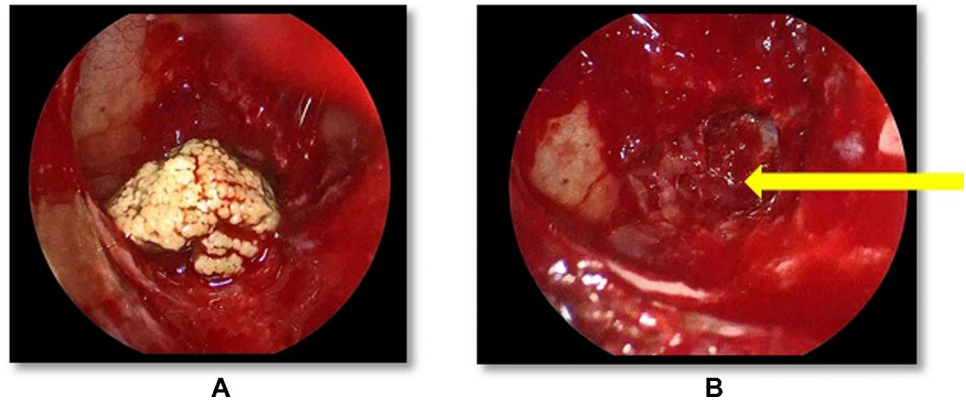


Fig. 2 (a) Axial and 2 (b) coronal post contrast Fat Sat T1 MRI demonstrating peripheral enhancement of mucocoele. (c) shows T2 Axial signal void in the mucocoele due to paramagnetic elements (iron and manganses) in the fungus

Fig. 3 (a) Intraoperative endoscopic images: a Cauliflower shaped Fungal ball, 3 (b) Clival osteomyelitis (arrow)



the mucocele drained and marsupialised. This then revealed endoscopic signs of clival osteomyelitis with exposed dura (Fig. 3). Specimens were sent for microbiology and frozen section histopathological assessment. Histopathology showed multiple nodules of eosinophilic material, which

were partly coated by neutrophil-rich fibrino purulent debris (Fig. 4a). The nodules comprised densely packed fungi and hyphae were identifiable within looser areas at the peripheries of nodules (Fig. 4b). Grocott histochemistry showed septate fungal hyphae branching at acute angles (Fig. 4c).

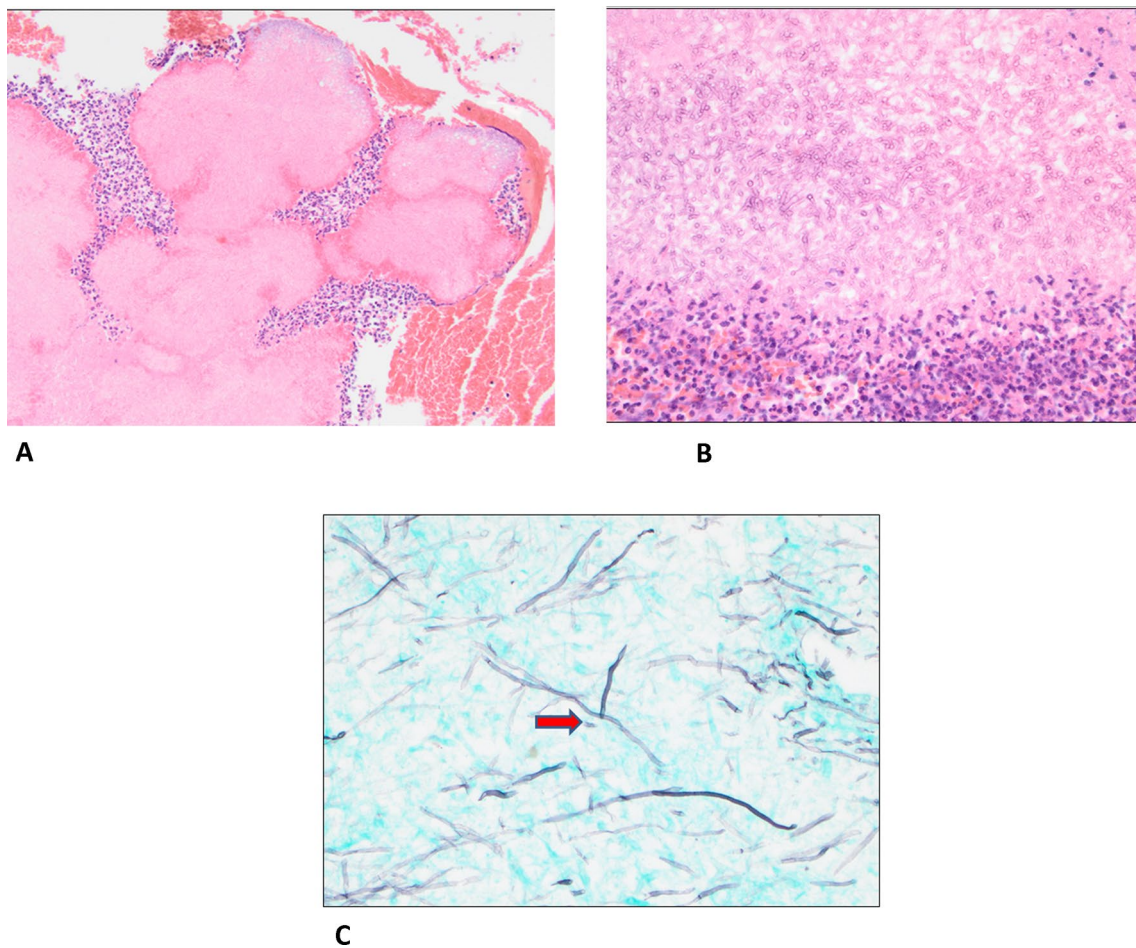


Fig. 4 (a) – H&E (×200). Multinodular aggregates of eosinophilic material partly surrounded by suppuration and blood clot. (b) – H&E (×400). Fungal hyphae identifiable within looser areas at the periph-

eries of nodules. (c) Grocott (×600). The fungi exhibit a branching septate pattern, with acute angles. This pattern is similar to that seen in the more common *Aspergillus* spp.

Minimal background mucus was of normal type, with no evidence of an allergic fungal sinusitis pattern. Mucosal fragments submitted with the mycetoma showed features of non-specific acute on chronic rhinosinusitis. Neither intramucosal/submucosal fungal elements nor neurovascular permeation were identified (Fig. 5) Primary culture of the tissue sample yielded fungal colonies which were identified using matrix-assisted laser desorption ionization-time of flight mass spectrometry (MALDI TOFF) as *Scedosporium* spp. This was formally confirmed as *Scedosporium apiopermum* by the mycology reference laboratory, Bristol which was sensitive to voriconazole and resistant to amphotericin B. This organism is known to be resistant to many antifungals. The patient was commenced on a 6-week course of voriconazole with close clinical and radiological surveillance. Post-operatively she was followed up for 12 weeks and is doing well and her presenting symptoms have resolved.

Discussion

Fungal infection is a rare complication of mucocoeles. The most common causative agent for fungal rhinosinusitis is *Aspergillus* [1]. Fungal infection mainly affects immunocompromised individuals. Isolated fungal sphenoiditis has a low incidence of 2%. *Scedosporium* spp. is found in soil and fresh water, especially stagnant or polluted water, throughout the world. It is mainly contracted from wood, hay or dogs.

Our patient was regularly involved in farm work and had a dog as a pet. It is acquired after inhalation of this organism into the lungs or paranasal sinus or after traumatic

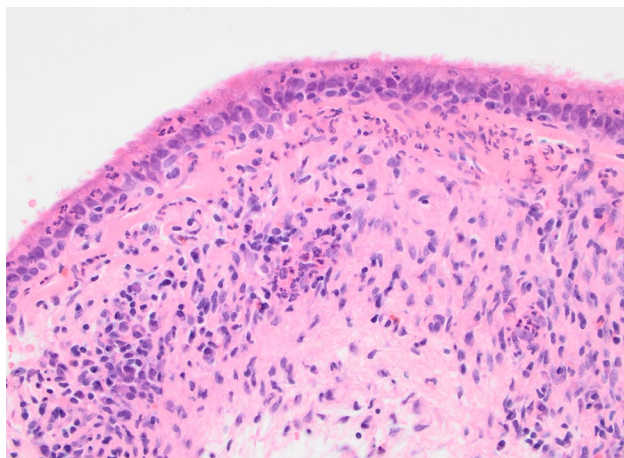


Fig. 5 H&E at $\times 200$ magnification. The sinonasal mucosa submitted with the mycetoma at frozen section shows non-specific acute-on-chronic rhinosinusitis. Neutrophils are present within the respiratory epithelium. The lamina propria contains a mixed inflammatory infiltrate that includes neutrophils, eosinophils, macrophages, lymphocytes, and plasma cells. No intramucosal/submucosal fungal elements were identified. There was no evidence of neurovascular permeation

inoculation through the skin [2]. It most commonly causes mycetomas in immunocompetent people and is seen mostly in temperate climates [3]. In immunocompromised patients, *Scedosporium* spp. are associated with sinusitis, invasive pneumonia and brain abscess. Central nervous system involvement is seen in near drowning patients.

Our patient was a 32 year old immunocompetent female with no co-morbidities or allergies. The diagnosis of a mucocele with mycetoma was unexpected.

Sphenoid sinus mucocele has varied presentations. Headache is the most common symptom. Bilateral visual disturbance is common and can be due to the involvement of Cranial Nerves II, III, IV and VI, presenting as decreased visual acuity, diplopia, blepharoptosis and external ophthalmoplegia [2, 4]. In our patient, unilateral eye symptoms were noted. Bony erosion of the clivus was noted radiologically suggestive of osteomyelitis which was confirmed intra operatively. However debridement of the bone was not possible as the erosions were up to dura, and bone samples was unavailable for histology. The osteomyelitis was a radiological diagnosis supported with clinical endoscopic evidence (Figs. 1, 2, and 3).

It is difficult to differentiate between *Aspergillus* and *Scedosporium* on histopathological assessment. Both show septate hyphae branching at acute angles (Fig. 4). Given that *Aspergillus* accounts for the majority of fungal rhinosinusitis cases, it may be tempting for the reporting pathologist to either suggest or support a diagnosis of *Aspergillus* infection based on H&E and fungal histochemical stains. However, this case illustrates the limitations of morphological assessment: microbiology was critical to establishing the diagnosis of *Scedosporium* infection, a distinction that is clinically significant due to the more aggressive course of *Scedosporium* and its resistance to certain antifungals. This case is informative to the reporting pathologist as it highlights the need for correlation with microbiological samples and the dangers of relying on morphological assessment of biopsy material. An erroneous assumption of *Aspergillus* infection could result in the administration of inappropriate/ineffective antifungal therapy and worse clinical outcome. There is only one other report in the literature of *Scedosporium apiopermum* infection in an immunocompetent patient [5].

Conclusion

We present a rare case of fungal sinusitis secondary to mucocele caused by *Scedosporium apiopermum* infection. The case highlights the difficulties of accurately diagnosing fungal sinusitis and the importance of microbiology given the morphological similarities of *Aspergillus* and rarer fungal species. We describe a triad of – headache, unilateral isolated sphenoid sinus opacification and absence of

immunosuppressive history. Considering the critical anatomical structures around the sphenoid sinus, we recommend examination under anaesthesia and surgical drainage for all patients with sphenoid sinus opacification and headaches. The osteomyelitic property of the pathogen can lead to life threatening complications including intradural abscess, meningitis and orbital apex syndrome. Our recommendation for post op management is 6 weeks of voriconazole.

When there are no bony erosion with 12 weeks of Voriconazole in cases of invasive disease or bony erosions. This should be closely followed up with full blood count and liver function tests in view of toxicity of voriconazole. Radiological follow up with MRI and CT scan is also essential.

Fungal disease has unpredictable presentations which warrant careful assessment and radical treatment.

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Compliance with ethical standards

Conflict of Interest None.

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