

# Isolated Sphenoid Aspergilloma: A Differential Diagnosis for Solitary Abducens Nerve Palsy

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## INTRODUCTION

Isolated sphenoid sinusitis (SS) is a very rare and potentially life-threatening occurrence. Because of its rarity, its true incidence is difficult to establish, but it lies within the range of 2.7% to 8% based on larger series (1–3). The incidence of isolated fungal disease is even lower. Diagnostically, SS represents a challenge as it does not present in a similar way to inflammation of the other paranasal sinuses, presenting symptoms being generally non-specific (4).

## Case Report

An 80-year old man was referred by his general practitioner to the ophthalmologist with diplopia of several weeks' duration. He was found to have a partial, non-resolving, right-sided abducens nerve palsy (ANP) on examination but no cause could be elicited clinically. His visual acuity and colour vision were normal. Magnetic resonance imaging (MRI) of the orbits and brain revealed a 1.5 x 2 x 1.5 cm well-encapsulated mass within the right side of a septate sphenoid air sinus (Fig. 1). The sinus was shown to be lined

by a thickened mucosa. The left sphenoid air cell and the rest of the paranasal sinuses were normal. The features of the mass suggested that of a benign tumour as the cause of the ANP. He was referred to the Ear Nose and Throat (ENT) Department for further management.

On further questioning by the ENT surgeons, he described mild nasal obstruction, bilateral rhinorrhoea and hyposmia. He denied any previous history of cerebrovascular accident, epistaxis or headaches and had never had prior sinonasal surgery. His medical history was significant for ischaemic heart disease for which he was on clopidogrel, and for which he had undergone coronary artery bypass surgery twelve years previously. Examination confirmed a right-sided ANP; nasal examination revealed a right-sided nasal septal spur, normal inferior and middle meati, and congestion of the right sphenoid ostium. The left sphenoid ostium was found to be healthy and the post-nasal space and head and neck examination were normal. Blood investigations performed at the time did not show evidence of granulomatous disease or diabetes mellitus. A decision to surgically remove the mass was made.

The patient underwent a right endoscopic transthemoidal sphenoidotomy. This revealed a brownish, calcified sphenoidolith, possibly of fungal origin. A provisional diagnosis of a fungus ball was made. This was confirmed after histopathological examination of the specimen (Fig. 2). A mass of

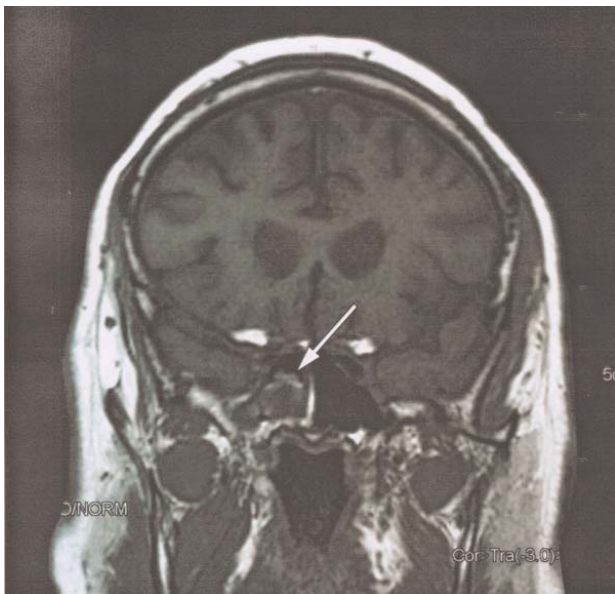


Fig. 1: Coronal MRI scan of the sinuses showing right sphenoid sinus aspergilloma (arrow).

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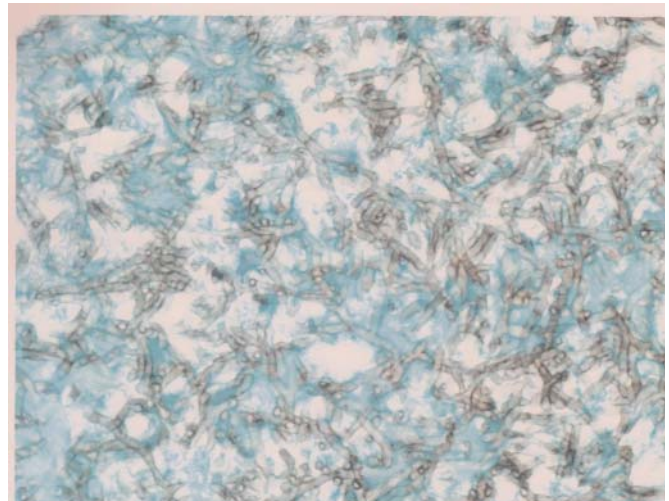


Fig. 2: Mass of broad, septate, branching hyphae, in keeping with *Aspergillus*, without intact mucosal tissue. Grocott stain. Magnification x 400.

fungal hyphae and mucus was demonstrated. The hyphae were broad, septate and branching in keeping with *Aspergillus*. A diagnosis of aspergilloma was thus made.

The patient was prescribed nasal saline douches and topical mometasone furoate and discharged from hospital the following day after an uneventful admission. On follow-up in the outpatient department one week later, he had begun to experience lessening of his diplopia. Three weeks after the surgery, he had made a full recovery from his ANP and there was no clinical evidence of disease recurrence on nasendoscopy.

## DISCUSSION

The non-specific presentation of Sphenoid Sinusitis (SS) can lead to a delay in diagnosis and lead to a potentially lethal situation (5). The presenting feature is often a complication of the sinusitis due to the proximity of the sinus to the orbit and the cavernous sinus (5). Abducens nerve palsy represents one of these complications and can serve as an early indicator for possible isolated sphenoid disease. Injury of the abducens nerve can occur anywhere along its long course, so the differential diagnosis of abducens nerve palsy demands thorough investigation to find the proper cause (6). As presented here, isolated fungal disease, including aspergilloma, is an important differential that is readily diagnosed through the use of computerized tomography and direct nasendoscopy (7) Magnetic resonance imaging may further define the extent and nature of the lesion (7).

The sphenoid sinus is in the geometric centre of the head and is absent only in 1% to 1.5% of the population (8). Because of its location, presenting symptoms during SS can be non-specific and optimal clinical examination is usually not possible due to its relative inaccessibility (4). The most commonly presenting complaints are headache and facial pain, but may also include rhinorrhoea and less frequently, nasal congestion (9). Headaches usually localize to the vertex of the skull (5). It is its anatomical relations, however, that are of interest to the physician. Superiorly, it is bounded by the pituitary gland, middle cranial fossa and the optic nerve and its chiasma; anteriorly by the nasopharynx, pterygoid canals and nerves, and the pteropalatine ganglion and artery; laterally by the cavernous sinus, internal carotid artery and cranial nerves III, IV, V<sub>1</sub>, V<sub>2</sub> and VI (10, 11). Because of its location deep within the skull base, infection can result in severe complications (12). Meningitis is the most common of these, but others include cavernous sinus thrombosis and orbital complications such as periorbital cellulitis, orbital cellulitis, subperiosteal abscess and orbital abscess (5).

Of the cranial nerves that can potentially be affected by SS, the abducens nerve is most likely (13). This is probably due to its more medial location in the cavernous sinus, and its variable course in which it may split into as many as five rootlets as it passes lateral to the intracavernous portion of the carotid artery (11, 14). It is thought that progressive thrombophlebitis affecting the inferior petrosal sinus leads to its

involvement during SS (15). This occurs because of a high local concentration of inflammatory mediators (3). Both unilateral and bilateral abducens palsy have been described, but the latter is much rarer (3).

The differential diagnosis of isolated sphenoid sinus disease is varied and includes SS, mucocele, fibrous dysplasia, meningoencephalocele, inverted papilloma, squamous cell carcinoma, liquor fistula, rhabdomyosarcoma, chordoma, carotid pseudoaneurysm and sphenochanal polyp, all of which may present with headache, visual disturbances and nasal congestion (5, 16, 17). Sphenoid sinusitis may be bacterial or fungal.

Fungal sinusitis is classified into invasive and non-invasive forms. Invasive forms may be acute (fulminant), chronic and granulomatous (indolent). Non-invasive forms include allergic fungal sinusitis (AFS) and fungus ball (mycetoma) (5, 18). Allergic fungal sinusitis is the most common form of fungal sinusitis (18). A paranasal sinus fungal ball is an extramucosal mycosis, usually occurring as a unilateral lesion, but multiple ones have been described (19, 20). They occur in elderly populations and tend to have a female predominance. Hormone changes which occur during this age might play a role in their pathogenesis (21). They carry a low morbidity and have a low recurrence rate (22). However, an immunocompromised state may cause this predominantly extramucosal disease to adopt an invasive form (23). They occur most commonly in the maxillary sinus, but can also occur in the sphenoid, ethmoid and frontal sinuses in decreasing frequency (19, 24). They are most frequently caused by *Aspergillus* and clinically they mimic chronic rhinosinusitis (25).

Diagnostic criteria are based on radiology and histopathology; fungal cultures are frequently negative (19, 25). Computerized tomography (CT) is of great help in the assessment of this disease, but occasionally MRI may be employed, especially when extensive skull base involvement is present (26). A CT finding of a heterogeneous sinus opacity with microcalcifications is very suggestive of the diagnosis but a homogeneous opacity may be encountered even with bone lysis (26). In the index patient, only a MRI was performed. This was in part due to the fact that he presented to the ophthalmologist first and sinus disease was not initially considered. Following discovery of sinus pathology and referral to ENT, further imaging via CT was not considered necessary as it was felt that no further useful information would be gained: the fungal ball was already clearly defined and the MRI provided clearer details about neurovascular structures within the cavernous sinus. In patients who present with an isolated ANP and no other symptoms, MRI may be considered as a first investigation. In one series of isolated, non-traumatic ANP, Brinnar *et al* demonstrated a wide spectrum of causes, including nasopharyngeal carcinoma, diabetes, cholesteatoma of the inner ear, carotid-cavernous fistula, subarachnoid bleeding, hydrocephalus, toxic ANP, multiple sclerosis, a clinically isolated syndrome suggestive of mul-

tiple sclerosis and Tolosa-Hunt syndrome. Based on the cases and a review of the literature, they argued that every patient with isolated, non-traumatic ANP requires a brain MRI as an initial diagnostic tool. If this finding remains inconclusive, additional tests including angiography and CSF examination should then be performed (6).

Surgical removal is the gold standard of treatment and several methods have been described, ranging from washouts through the anterior sinus wall or ostium to intranasal sphenoidotomy to trans-septal, transantral or transthemoidal sphenoidectomy (27). An endoscopic approach, however, is considered to be the most reliable and safest one (19, 28). It involves opening the infected sinus cavity at the level of its ostium and removing fungal concretions while sparing the normal mucosa (28). The index patient underwent endoscopic transthemoidal sphenoidotomy because of surgeon familiarity with the approach. Treatment of this disease is considered to be primarily surgical; postoperative antifungal therapy is not indicated (20, 28).

In conclusion, due to the complex anatomy of the abducens nerve, the variety of pathology that may result in its palsy and the non-specific nature in which isolated sphenoid sinus disease presents, isolated ANP can be very difficult to diagnose clinically and a wide differential diagnosis should be considered. Isolated aspergilloma remains an important differential as it represents one that may result in significant morbidity, but is readily amenable to surgical intervention with full recovery of the palsy expected if discovered early. Computed Tomography and MRI remain important modalities in its diagnosis and planning of further management.

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