Onodi Cell Lymphoma with Secondary Fungal Sinusitis presenting as Orbital Apex Syndrome

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Abstract

A 53-year-old Chinese gentleman with underlying diabetes mellitus presented with one week history of right painless blurring of vision and ptosis with a visual acuity of 6/24. Examination suggested presence of orbital apex syndrome. Imaging revealed right orbital apex compression by fluid filled sphenoidonal air cell (onodi air cell) with soft tissue mass in the right maxillary sinus and nasal cavity and mucosal thickening in the air sinuses. Patient underwent functional endoscopic sinus surgery and septoplasty. Thick pus secretions in the sphenoidal sinus grew Aspergillus fumigatus and biopsy of the polypoidal mucosa filling up osteomeatal complex, maxillary antrum, ethmoid and onodi air cell revealed Natural Killer (NK)/T-cell lymphoma of nasal type positive for CD3, CD30 and CD56 marker as well as Epstein-Barr virus stain. His vision remained the same post surgery. Patient was started on intravenous Amphotericin B and referred to the Hematology team for initiation of chemotherapy however he later succumbed to his illness.

Keywords: Onodi Cell; Lymphoma; Sinusitis; Orbital Apex Syndrome

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Introduction

A sphenoidonal air cell (Onodi cell) is an anatomic variant of the paranasal sinus. It is a pneumatized posterior ethmoid air cell that extends into the sphenoid sinus and may be associated with aeration of the optic strut and anterior clinoid process. Inflammation of these air cells can lead to the formation of mucocele, with subsequent pressure on surrounding structures [1].

Although rare; due to its relative position to the optic nerve and orbital apex, Onodi cell sinusitis may lead to compressive optic neuropathy or orbital apex syndrome. The development of an Onodi cell mucocele and subsequent effect on the orbit is a rarely reported but important phenomenon [1-2]. Here, we would like to report one such case of lymphoma affecting the Onodi cell with secondary fungal sinusitis which presented as orbital apex syndrome without prior symptoms suggestive of Otorhinolaryngology (ORL) pathology.
Case Report

A 53-year-old Chinese gentleman, who was newly diagnosed with diabetes mellitus, presented to us with one week history of acute, painless blurring of vision and complete ptosis of the right eye. He had no prior history of fever, headaches, upper respiratory tract infection or trauma.

On examination, his best corrected visual acuity was 6/24 on his right eye and 6/18 on his left eye. He had a right relative afferent pupillary defect, with ptosis and limitation of superior, medial and inferior gaze. Pupils were both 3mm, round and reactive. Fundus examination showed right optic disc swelling with peripapillary splinter hemorrhages and hard exudates at the macula. No other neurological abnormalities were seen. Contrast-Enhanced Computed Tomography (CECT) of brain, orbit and paranasal sinuses was done and revealed optic nerve thickening with compression of the right orbital apex by Onodi air cell sinusitis, with soft tissue mass in the right maxillary sinus and nasal cavity and mucosal thickening in the air sinuses cells (Figure 1).

Patient underwent a functional endoscopic sinus surgery and septonplasty. Polypoidal mucosa was seen filling up osteomeatal complex, maxillary antrum, ethmoidal and Onodi air cells. Thick pus secretions seen in the sphenoidal sinus were removed and sent for culture, which grew *Aspergillus fumigatus*. Polypoidal mucosa of the osteomeatal complex, maxillary antrum, ethmoidal and onodi air sinuses cells were biopsied for Histopathological Examination (HPE) which revealed Natural Killer (NK)/T-cell lymphoma of nasal type.

There were small to medium sized cells seen with irregular membrane and small inconspicuous nucleoli on HPE. Extensive areas of necrosis with numerous apoptotic bodies were observed. Angiocentricity was present. The mitotic figures and atypical cells were positive for CD3, CD30 and CD56. Focal cells were also positive for Epstein-Barr virus (EBV) immunohistochemistry stain. Stains for granzyme B, perforin and EBV-encoded RNA were not done as they are not available at our center. There was no evidence of *Aspergillus fumigatus* hyphae infiltrating the tumour cells directly on histopathology. Patient was started on intravenous Amphotericin B and was referred to our Hematology team for initiation of chemotherapy. His vision however remained the same post-surgery and patient succumbed to his illness shortly thereafter.

Discussion

An Onodi cell is the most posterior ethmoid cell, which pneumatizes the sphenoid sinus laterally and superiorly [3]. It is closely proximal to the optic nerve and orbital apex and an optic canal bulge can be seen endoscopically [3]. The incidence of Onodi cells are 8-14% [4].

Mucocoeles are usually secondary to an obstruction in the main ostium of the sinuses due to inflammation, trauma, surgery or tumour. Chronic expansion of the mucocele causes pressure within the sinus to build up leading to bone expansion or erosion with pressure on the surrounding structures. This may induce a direct mechanical compression onto the optic nerve and orbital apex; causing compressive optic neuropathy or orbital apex syndrome [5]. The visual loss in our patient was thought to be because of compressive optic neuropathy rather than an invasion of the optic nerve by T-cell lymphoma as there was no evidence of optic nerve invasion seen on the CECT scan. We believe that the tumour which appeared as polypoidal mucosa most likely was localized at the osteomeatal complex and extended into maxillary, sphenoidal and onodi air sinuses as the T-cell lymphoma was of the nasal type on HPE. Neuroimaging is essential in its detection and differentiation from an underlying tumor is aided by the addition of an MRI examination preoperatively [6]. In our patient, it is presumed that the sinus involvement on the CECT scan indicated both T-cell lymphoma and Aspergillus as there was a soft tissue mass seen in the right maxillary sinus and nasal cavity as well as mucosal thickening in the air sinuses.

Although an identifiable cause of mucocele in Onodi cell sinusitis are not found in most cases, histopathological examination of the air sinuses cell biopsies of our patient revealed nasal-type NK/T-cell lymphoma. Patients usually present with nasal obstruction, or purulent or bloody
rhinorrhoea; although these and other associated symptoms were not present in our patient. This lymphoma is usually associated with mucosal ulceration [7].

It is probable that our patient developed *Aspergillus* fungal sinusitis secondary from these underlying malignant lesions, compounded with the fact that he was newly diagnosed with diabetes mellitus which rendered him immunocompromised. This fungal infection was most likely still in its early stages when detected as there was absence of the usual headache and facial pain which are normally out of proportion to clinical evaluation and imaging findings [8]. There were also no associated ulcerations of nasal or oral mucosa, necrosis or eschar formations which are all specific clinical signs in fungal sinusitis [8].

Early recognition and treatment in cases of optic neuropathy with associated loss of vision in patients with mucoceles is important [9]. Prognosis of visual acuity recovery is related to the time interval between onset of symptoms and surgery, stating one month of delayed treatment as a cut-off for poorer outcome [10]. Thus surgical decompression should be attempted in all such patients for therapeutic and diagnostic purposes.

In conclusion, Onodi cell mucocele, although rare should be considered as an etiology of orbital apex syndrome. Treatment of orbital apex syndrome usually requires a multidisciplinary approach involving the Ophthalmic and ORL team. Visual function recovery may be achieved with prompt treatment and intervention.

References

**Figure 1** Axial CT imaging showing fluid filled onodi cell with mucosal thickening compressing the right orbital apex