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Sphenoethmoidal air cell sinusitis: A rare cause of recurrent optic neuritis

Wei-Lin OuYang [a], Christopher Long [b], Saif Azam [a], Cassie Jia [a], Kimberly Gokoffski [b], Brandon Wong [b], Vishal Patel [a,c,*]

[a] Department of Radiology, Keck School of Medicine, University of Southern California, 1500 San Pablo Street, 2nd Floor, Los Angeles, CA, 90033, USA
[b] Department of Ophthalmology, Keck School of Medicine, University of Southern California, 1450 San Pablo Street, Los Angeles, CA, 90033, USA
[c] Mark and Mary Stevens Neuroimaging and Informatics Institute, University of Southern California, 2025 Zonal Avenue, Los Angeles, CA, 90033, USA

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ABSTRACT

Purpose: To describe the clinical presentation, imaging characteristics and management of a rare case of Onodi cell sinusitis associated with optic neuropathy.

Observations: A 46-year-old male presented to the emergency department with progressive left eye vision loss over the course of ten days. The constellation of findings from this patient's history, physical, and fundoscopic exam, as well as CT and MR imaging led to the diagnosis of Onodi air cell sinusitis complicated by optic neuropathy. The patient's symptoms resolved fully and vision returned to baseline with oral antibiotics.

Conclusions and Importance: The sphenoethmoidal air cell, also known as the Onodi cell, is an anatomic variant of the paranasal sinuses whose spatial relationship with important neurovascular structures carries significant clinical implications when it becomes inflamed or infected. Our case of Onodi cell sinusitis complicated by optic neuropathy demonstrates how vision loss secondary to sinusitis may resolve with oral antibiotic treatment. We additionally review the relevant anatomy, clinical course and treatment of Onodi cell pathologies.

1. Introduction

Four groups of paranasal sinuses border the orbit. The maxillary sinuses are the largest and located inferior to the orbital floor. The frontal sinuses are located superior to the orbits and ethmoid sinuses, and they border the medial portion of the orbital roof. The sphenoid sinuses are located posterior to the ethmoid sinuses; in the conventional configuration, their lateral walls may border the orbital apex, the optic canal and the inferior orbital fissure.[1,2]

Lastly, the ethmoid sinuses are highly variable in their configuration and formed by multiple air cells: 3 to 4 cells at birth and 10 to 15 cells by adulthood.[3] They are located inferior to the frontal sinuses and anterior to the sphenoid sinuses. They border the medial aspect of the orbit, from which they are separated by the lamina papyracea. They are divided into the anterior and posterior ethmoid groups by the basal lamella of the middle turbinate, with the anterior portion inserting vertically into the crista ethmoidalis and the posterior portion attaching to the lamina papyracea horizontally.[2,3]

There are two primary drainage pathways of the paranasal sinuses. Anteriorly, the osteomeatal complex drains the frontal sinuses via the frontal recess and anterior ethmoid sinuses and maxillary sinuses via the ethmoid infundibulum into the middle meatus. Posteriorly, the sphenoethmoidal recess drains the posterior ethmoid sinuses and the sphenoid sinuses into the superior meatus.

Pathological processes involving the paranasal sinuses are prevalent, with acute and chronic sinusitis affecting 1 out of 7 adults in the United States and accounting for approximately 16 million office visits per year.[4] Given the intimate relationship between the paranasal sinuses and the orbits, any pathological process involving the sinuses has the potential to progress into the orbits. Moreover, anatomical variants of the paranasal sinuses may predispose patients to sinus pathologies.

The sphenoethmoidal air cell, also known as the Onodi cell, is an anatomical variant of the most posterior ethmoid cell which extends posteriorly along the superior and lateral margins of the sphenoid sinus. Notably, it may lie in close proximity to the intracanalicular portion of the optic nerve and the cavernous sinus, and its relationship to these neurovascular structures carries clinically significant implications.

We present a case of Onodi cell sinusitis complicated by optic neuropathy and additionally review the relevant anatomy, clinical course and treatment of Onodi cell pathologies.

* Corresponding author. Department of Radiology, Keck School of Medicine, University of Southern California, 1500 San Pablo Street, 2nd Floor, Los Angeles, CA, 90033, USA.
E-mail address: vishal.patel@med.usc.edu (V. Patel).

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2. Case report

A 46-year-old male with history of chronic sinusitis status post right maxillary antrostomy presented to the emergency department with progressive left eye vision loss over the course of 10 days. At initial symptom onset, he reported retroorbital pressure-like pain and discomfort with mild tenderness to palpation, but this resolved prior to presentation. The patient reported a history of similar symptoms at least 3 times over the previous 3 years, each resolved with antibiotic treatment.

At time of presentation, visual acuity (VA) was 20/20 on the right, count fingers on the left. Pressures were normal. An afferent pupillary defect (APD) was noted on the left. Color plate testing and red desaturation were normal on the right, but limited by VA on the left. Light saturation was 100% on the right, 1–2% on the left. Slit lamp exam was within normal limits. On dilated fundus exam the disc was sharp and pink on the right while there was evidence of 360° disc edema with small vessel obscuration on the left.

An MRI of the orbit with and without contrast was requested and demonstrated thickened, enhancing mucosa along the superolateral wall of a left Onodi cell which extended into the left anterior clinoid process and surrounded the optic canal, as well as linear enhancement along the optic nerve sheath (Fig. 1). A CT of the paranasal sinuses obtained during the current episode confirmed mucosal thickening and opacification of the previously aerated left Onodi cell and anterior clinoid process, and it additionally identified an area of focal osseous dehiscence along the lateral aspect of the Onodi cell that established continuity between the sinus and the orbital apex (Fig. 2).

The patient was admitted briefly to the neurology service for evaluation of optic neuritis, with ophthalmology and otolaryngology services consulting. Erythrocyte sedimentation rate and C-reactive protein levels were normal, antinuclear and perinuclear anti-neutrophil cytoplasmic antibodies were not detected, and screening studies for Bartonella, tuberculosis, and syphilis were negative. A trial of oral amoxicillin-clavulanate 875mg–125mg twice daily, loratadine 10 mg daily, fluticasone nasal spray 100 mcg twice daily and saline nasal irrigation resulted in improvement of visual symptoms after 48 hours. He was therefore discharged to complete a 10-day course of oral antibiotic therapy. At his 2 week follow up visit, marked improvement was noted in the patient's exam, with VA on the left improved to 20/40, 20/30 with pinhole. There was no APD. Color plates were full on the right, 8/10 on the left. Fundus exam was notable for improved edema, sharpened disc margins and decreased small vessel obscuration.

Complete resolution of symptoms was noted at follow up 2 months after diagnosis. VA on the left was 20/25 and color plates were full. On fundus exam, the left disc was sharp without any vessel obscuration, indicating the disc edema had resolved. The constellation of findings from the patient's history, fundoscopic exam, imaging and the positive response to antibiotic therapy supported the diagnosis of Onodi air cell sinusitis complicated by optic neuropathy.

3. Discussion

The optic nerve can be divided into intraocular, intraorbital, intracanalicular and intracranial portions. It extends from the posterior globe through the orbit and the optic canal, which is adjacent to the sphenoid bone, before emerging into the suprasellar cistern and terminating at the optic chiasm. There are multiple etiologies of optic neuropathy, including but not limited to demyelination, ischemia, infiltration, compression, toxicity, trauma and paraneoplastic disease. The clinical history, including the acuity and pattern of visual symptom onset, physical and fundoscopic exams, and supportive findings from radiologic workup are paramount in identifying the correct cause of optic neuropathy.[5]

This case demonstrated a rare instance of secondary optic neuropathy from adjacent Onodi cell sinusitis. The Onodi cell was first described in 1904 by Adolf Onodi as an anatomical variant of the most posterior ethmoid air cell that extends along the superior and lateral border of the sphenoid sinus, near the intracanalicular portion of the optic nerve with only a thin sliver of bone to separate these structures.

Generally, Onodi cells are asymptomatic unless they are complicated by sinus disease, which may be more prevalent in those who possess the variant.[6] Reported etiologies of Onodi cell sinusitis may include infection by bacteria or fungi[7] and inflammatory changes from compressive lesions such as polyps,[8] mucoceles[9,10] and even necrotic tumor.[11] These disease processes can secondarily involve the optic nerve, and early diagnosis and treatment can result in improvement of visual symptoms.[7,8,10]

Other reports have described cases of Onodi cell sinusitis complicated by abscess formation and invasive mucormycosis eventually leading to orbital apex syndrome,[7,12] a more catastrophic ophthalmologic condition characterized by acute vision and ophthalmoplegia. After the diagnosis was confirmed in these cases, patients were treated with surgical decompression and appropriate antimicrobial therapy and went on to experience resolution of acute infectious and visual symptoms. However, if delays in diagnosis or treatment occur, as in the case of one reported individual with mucocele-associated Onodi sinusitis and another with chronic osteomyelitis, optic nerve atrophy and blindness may persist even after surgical intervention and medical treatment.[9,11]

CT scan is considered the best initial radiographic evaluation of the paranasal sinuses and the surrounding structures. The incidence of the Onodi cell detected by CT scan ranges from 8 to 30%, with an increased prevalence in the Asian population.[13,14] On imaging, it may be difficult to distinguish the Onodi cell from the sphenoid sinus or an overriding posterior ethmoid cell. However, failure to do so may lead to misdiagnosis, incomplete surgical or medical treatment, and risk of permanent

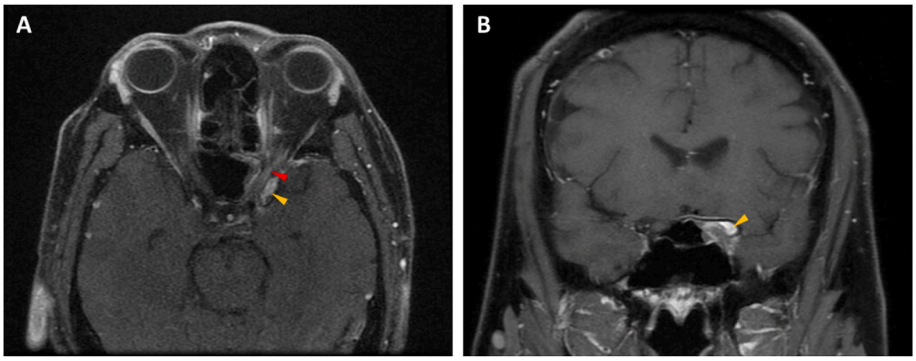

Fig. 1. Axial (A) and coronal (B) T1-weighted, post-contrast, fat-suppressed MR images of the orbits obtained at presentation. There is thickened, enhancing mucosa in a left Onodi cell, extending into the anterior clinoid process (yellow arrowheads). Also seen is linear enhancement along the optic nerve sheath within the optic canal (red arrowhead). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

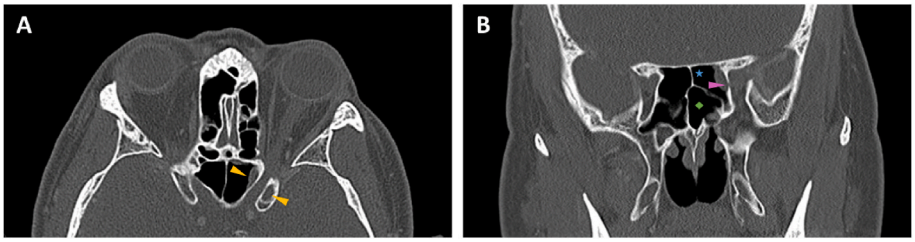

Fig. 2. Axial (A) and coronal (B) CT images of the paranasal sinuses show a left Onodi cell (blue star) superior to the sphenoid sinus (green diamond). Mucosal thickening in the Onodi cell and the contiguous pneumatized anterior clinoid process surrounds the optic canal (yellow arrowheads). Also seen is a site of focal osseous dehiscence along the lateral aspect of the Onodi cell that establishes continuity with the orbital apex (purple arrowhead). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

optic nerve damage.[14] Distinguishing features of the Onodi cell are its lateral and superior pneumatization relative to the sphenoid sinus, its close association with the optic canal and, in some cases, prominence of the optic nerve tubercle and internal carotid artery.[15] MR imaging of the orbit has also proven useful in further characterizing the involvement of surrounding neurovascular structures, as in this and several other cases.[7,9–12]

There is currently no consensus in the management of pathologies involving the Onodi cell, as it depends significantly on the severity of symptoms. In patients with symptoms lasting longer than one month whose CTs demonstrate compressive optic neuropathy due to a Onodi cell mucocele, the most favored treatment is surgical decompression with adjuvant corticosteroid.[16,17] In our case, there was no compressive lesion, and conservative management with antibiotic therapy likely led to decreased inflammation within the Onodi cell and optic nerve sheath within the adjacent optic canal.

4. Conclusion

In patients presenting with optic neuropathy, adequate history, physical exam and supportive imaging findings are essential to achieve accurate diagnosis and management. It is important for radiologists and ophthalmologists to be familiar with the Onodi cell given its anatomical location, association with sinusitis and implications for treatment. Thorough radiographic evaluation of the optic nerve is warranted when there are image findings of Onodi cell-associated pathologies. Early diagnosis, medical therapy and surgical intervention, when indicated, can lead to resolution of symptoms in patients suffering from this treatable cause of acute vision loss.

Patient consent

Consent to publish this case report has been obtained from the patient in writing.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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