Compressive optic neuropathy due to a large Onodi air cell: A case report and literature review

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ABSTRACT

Background: *To report an unusual case of compressive optic neuropathy secondary to a large onodi air cell.*

Method: *Case report.*

Results: *A 50 year-old gentlemen presented to the eye clinic with left eye painless loss of vision for one day. Visual acuity was counting finger in the left eye with a positive relative afferent pupillary defect (RAPD). Dilated left fundus examination revealed a pale optic disc. A computed tomography of orbit and brain showed a large left sphenoid sinus with onodi-cell-like projection on the left superior margin of left optic canal impinging on the left optic nerve. He was referred to the otorhinolaryngology team and subsequently underwent left optic nerve decompression. Post-operatively, his left visual acuity improved to 6/60 with reversal of RAPD.*

Conclusion: *There are many causes of optic neuropathy and compressive optic neuropathy due to large onodi air cell is uncommon. Acute unilateral loss of vision heralds from a multitude of sinister causes and junior residents should be vigilant that onodi air cell pneumotisation could be one of them.*

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The paranasal sinus possesses an interface with the orbit, which forms its walls. The frontal sinus forms the medial part of the orbital roof, whereas the ethmoid sinus is separated from the medial wall of the orbit by the lamina papyracea. The maxillary sinus constitutes the floor of the orbit, and the sphenoid sinus lies within the body of the sphenoid bone,[1] which surrounds the orbital apex. This close anatomic relationship is of paramount clinical significance whereby any pathologic process of the paranasal sinus can affect the adjacent orbit. Feasible mechanisms of optic neuropathy could be attributed to the mass effect of direct compressive impact; ischemic insult due to the compromised blood supply of the optic nerve, with eventual development of optic atrophy; or spread of suppurative focus from the adjacent paranasal sinus into the orbital cavity.[2] The posterior paranasal sinus (specifically, the posterior ethmoid and sphenoid sinus) pathology is commonly associated with ophthalmic manifestations, such as blurring of vision or ophthalmoplegia.[2] The sphenoethmoidal or Onodi air cell is essentially an anatomic variant of a posterior ethmoidal air cell, which may pneumatize posteriorly into the sphenoid sinus.[3] Although it prevails radiographically on computed tomographies (CT) in only 7% of the population, this sphenoethmoidal complex can actually be found in 39% of anatomic dissections.[4] We herein present an uncommon case of retrobulbar compressive optic neuropathy secondary to a large Onodi air cell in a patient with no known previous nasal pathology, with visual improvement after endoscopic decompression.

CASE REPORT

A 50-year-old laborer presented to the eye department with complaints of a sudden onset of painless loss of vision in his left eye, which was preceded by a low-grade fever 2 weeks earlier. He noticed the acute loss of vision on waking up in the morning. Apart from the absence of other ocular or nasal symptoms, there also was no headache or other neurologic deficits. He had no known underlying medical illnesses. Besides no history of sinusitis or nasal surgical procedures, he also had no history of prior trauma. Results of an ophthalmic examination revealed left visual acuity of counting fingers, with a positive relative afferent pupillary defect. Anterior segment examination of the left eye was normal. A fundoscopy revealed a pale left optic disc with a cup-to-disc ratio of 0.4. The remainder of the ophthalmic examination was unremarkable, with no optic disc swelling or intraretinal hemorrhages. The retina and macula were flat, with no striaes seen. Also, the retinal vessels appeared normal. The fellow eye was normal, with a best corrected visual acuity of 6/6. The intraocular pressures in both eyes were within the normal range. The rest of the cranial nerves function was intact as well.

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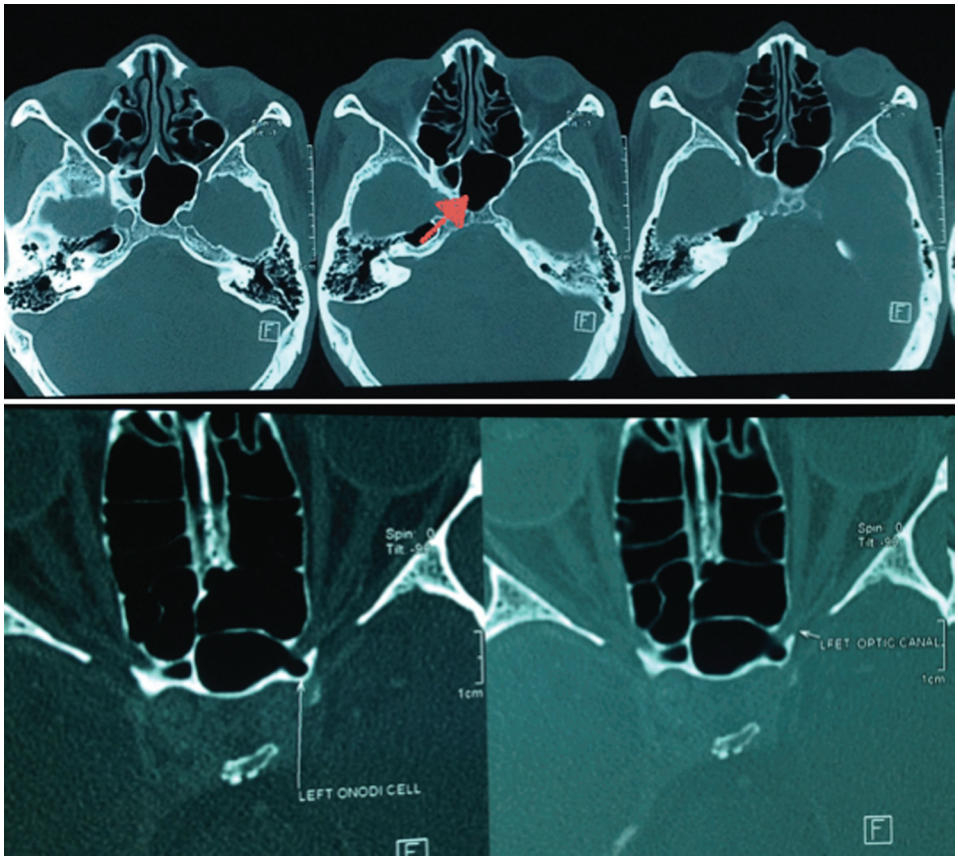

Figure 1. *Large left sphenoidal sinus with Onodi cell-like projections (above, red arrow) on the left optic canal (below, left); Left Onodi air cell (white arrow) impinging on the left optic nerve.*

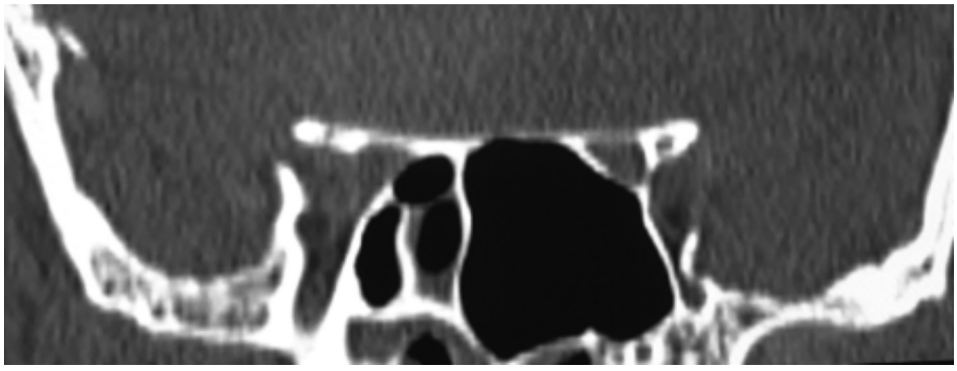

Figure 2. *Superior wall of the left sphenoid sinus dehiscence.*

A preliminary diagnosis of optic neuropathy was made, and further investigations were conducted to determine the cause. Routine blood investigations were noncontributory. An axial plane of contrasted CT of the orbit demonstrated a big left sphenoid sinus (Fig. 1) with Onodi-cell–like projection on the left superior margin of the left optic canal, which impinged on the left optic nerve (Fig. 1). The coronal plane of the CT orbit (Fig. 2) revealed thinning of the superior wall of the left sphenoid sinus adjacent to the left optic nerve. There was no retro-orbital mass. Retro-orbital fat appeared normal, so did the optic chiasm. The bilateral extraocular muscles were symmetrical. There was no sinusitis seen on the CT. The brain imaging results were normal.

Based on the diagnosis of compressive optic neuropathy secondary to the large Onodi air cell, he was referred to the otolaryngology team. He, subsequently, underwent a left optic nerve decompression 6 weeks after his initial presentation. During surgery, his left middle turbinate was lateralized and the left maxillary ostium mucosa was debrided. His osteomeatal complex and ethmoid cells were noted to be normal during surgery. The medial part of the optic nerve wall was opened (Fig. 3), and surgery proceeded without any complications. After surgery, he was commenced on oral steroid, and, 5 months later, his left vision improved to 6/60 with the reversal of relative afferent pupillary defect.

DISCUSSION

The paranasal sinus possesses an interface with the orbit, which forms its walls. The frontal sinus forms the

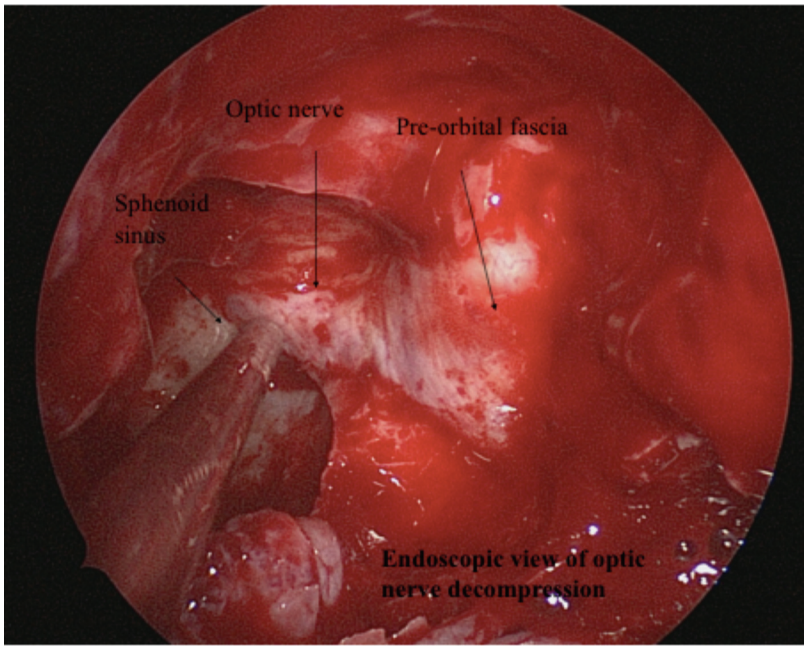

Figure 3. *Intraoperative endoscopic view of left optic nerve decompression.*

medial part of the orbital roof whereas the ethmoid sinus is separated from the medial wall of the orbit by the lamina papyracea. The maxillary sinus constitutes the floor of the orbit, and the sphenoid sinus lies within the body of sphenoid bone,[1] which surrounds the orbital apex. This close anatomic relationship has paramount clinical significance whereby any pathological process of the paranasal sinus can affect the adjacent orbit. Feasible mechanisms of optic neuropathy could be attributed to the mass effect of direct compressive impact; ischemic insult due to the compromised blood supply of the optic nerve, with eventual development of optic atrophy; or spread of suppurative focus from the adjacent paranasal sinus into the orbital cavity.[2] The posterior paranasal sinus (specifically the posterior ethmoid and sphenoid sinus) pathology is commonly associated with ophthalmic manifestations, such as blurring of vision or ophthalmoplegia.[2]

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Optic neuropathy has a myriad of causes. The common causes of compressive optic neuropathy include orbital and intracranial meningiomas, pituitary adenomas, and intracranial aneurysms.[5] Optic neuropathy is rarely caused by an Onodi air cell. The lateral or superior pneumatization of this posterior ethmoidal air cell into the sphenoid sinus[3] may directly abut the optic nerve.[6] This is clinically significant because any Onodi air cell pathology may affect this cranial nerve in view of their close anatomic proximity. Likewise, any sphenoid sinus or posterior ethmoidal complex surgeries and instrumentations may also injure the optic nerve.[3]

A literature review showed few reports of Onodi cell mucoceles[7–10] and polyps[11] that result in optic neuropathies. Indeed, only one case of orbital apex syndrome secondary to Onodi cell mucocele has been reported to date.[12] Therefore, we are of the impression that our case could potentially be the first case of a mere large-sized Onodi air cell itself, which resulted in compressive optic neuropathy, evidenced by the absence of other brain, sinus, or orbital pathologies on neuroimaging. Because the aforementioned presentation was his first visit to a tertiary center, our patient did not have any previous head or paranasal CTs for comparison. Although we believed that the left Onodi air cell was acutely pneumatized, which correlated to the sudden visual loss symptom, we were unable to reaffirm that hypothesis. The degree of sphenoid sinus pneumatization varies individually.[13] DeLano *et al.*[6] stated that "the greater aeration of the sinus and surrounding structures, the greater likelihood of optic nerve or canal indentation on sphenoidal sinus and of bone dehiscence."

A CT, specifically of the coronal plane, is an excellent diagnostic tool of choice to accurately demonstrate the relationship among the optic nerve and the paranasal sinuses, the osteomeatal complex, the course of the optic nerve, and the optic foramen.[6] Axial imaging is used to plan functional endoscopic sinus surgery in diseased posterior ethmoidal and sphenoidal sinuses.[6] Interestingly, with particular regard to our patient, his axial plane demonstrated the Onodi air cell more

prominently than the coronal plane in the absence of sinusitis, mucocele, or polyp.

The management of an Onodi air cell remains debatable, although surgical decompression prevails to be the mainstay of the management, with adjuvant systemic corticosteroids. Wu *et al*.[8] believed that surgical intervention should be attempted for all patients with optic neuropathy due to Onodi cell mucocele. The endoscopic transnasal decompression approach is less invasive, with fewer complication rates compared with the transcranial approach,[11] which makes the former a more-preferable method. Early surgical intervention to decompress the optic nerve may possibly reverse,[7] if not preserve, the visual functions of the optic nerve. Poor visual prognostic factors include poor presenting visual acuity and the sudden onset of symptoms.[7] The interval from the onset of symptoms to surgical intervention is also crucial for visual recovery. Nonake *et al*.[7] indicated that surgery should be performed within 24 hours from the onset of visual loss and that the visual potential will be poor if intervention occurs 1 month after presentation.

CONCLUSION

Paranasal sinus diseases often present with ophthalmic symptoms.[2] Because trainees and junior residents are the first clinicians to encounter patients with such ailments, it is of utmost importance that they are aware of the Onodi air cell and its implications on the optic nerve. A high level of suspicion is entailed for a correct diagnosis because prompt referral by the ophthalmology team, timely comanagement, and surgical intervention by the otorhinolaryngology team could potentially reverse permanent damage to the optic nerve, as illustrated by this case. Acute unilateral loss of vision heralds from a multitude of sinister causes, and junior residents should be vigilant that Onodi air cell pneumatization could be one of them.

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