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Optic neuritis caused by aspergilloma within Onodi cell pyomucocele in a 62-year-old woman

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ABSTRACT

Onodi cells are anatomical variants of the posterior ethmoidal cells, which are greatly pneumatized laterally, and to some degree superiorly, to the sphenoid sinuses. The clinical importance of the Onodi cell is that it contains the optic canal and lies in close proximity to the optic nerve. Although the incidence of Onodi cell pathologies such as mucocele is extremely low, they may cause ophthalmological complications due to the close anatomical proximity between the Onodi cell and the optic nerve. Optic neuritis caused by aspergillosis of the Onodi cell, presenting in the Emergency Department as an aspergilloma within a pyomucocele, has not been reported previously. Here, we describe the first such case; the aspergilloma within the pyomucocele was completely removed via surgical endoscopy.

Key words: optic neuritis, onodi cell pyomucocele, aspergilloma, CT, MRI

INTRODUCTION

Onodi cells are anatomical variants of the posterior ethmoidal cells, which are greatly pneumatized laterally, and to some degree superiorly, to the sphenoid sinuses. (1) The clinical importance of the Onodi cell is that it contains the optic canal and lies in close proximity to the optic nerve, as a result of which Onodi cell pathologies may lead to ophthalmological complications. (2) Although cases of isolated mucoceles in Onodi cells have been reported, there is only one case report of an aspergilloma within a simple mucocele causing compressive optic neuropathy. (3) However, there is no reported case of optic neuritis caused by an aspergilloma within a pyomucocele in an Onodi cell. Here, we present the first reported case, which was initially diagnosed in the Emergency Department.

CASE PRESENTATION

A 62-year-old woman was admitted to the Emergency Department with aggravated headache. The onset of the headache was 1 week previously and was accompanied by mild fever and visual dimness of the right eye. The patient denied a history of cardiovascular disease, stroke, or connective tissue disease. Her past medical history included diabetes mellitus, diagnosed 5 years previously. Review of vital signs revealed

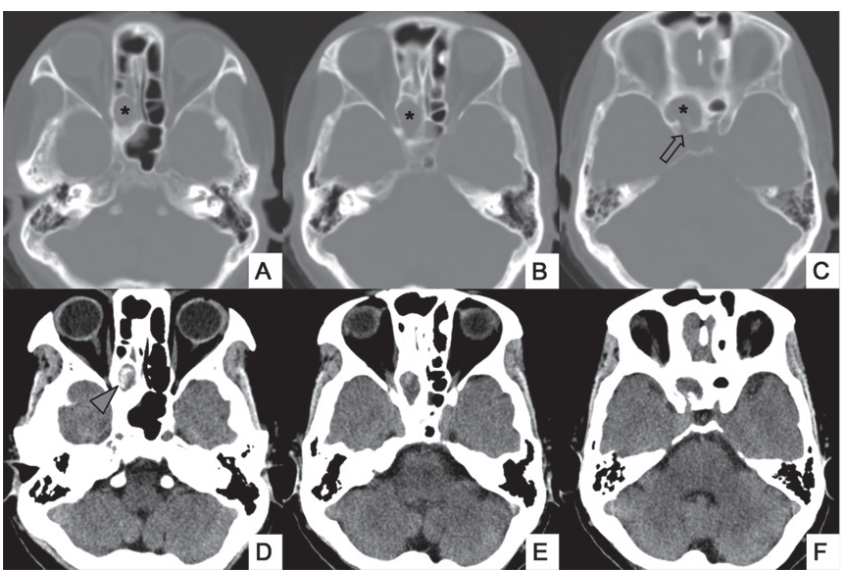

Figure 1.
A-C. Serial brain computed tomography (CT) images with bone setting show posterior ethmoidal cells (asterisks) greatly pneumatized far laterally, indicating an Onodi cell. Optic canal (arrow) within Onodi cell is seen.
D-F. Corresponding brain CT images with soft tissue setting show low-attenuation (approximately 4-5 HU) and expansile cystic mass with internal punctate calcification (arrowhead), presumed to be an aspergilloma within a mucocele.

blood pressure of 113/81 mm Hg, pulse of 56 beats/minute, respiration rate of 18 breaths/minute, and body temperature of 37.5°C. Results of laboratory tests were within the normal range except for serum glucose (133 mg/dL), white blood cell count (13,900 cells/ L), erythrocyte sedimentation rate (69 mm/h), and C-reactive protein (5.70 mg/dL).

Initially, we performed computed tomography (CT) of the brain to exclude intracranial hemorrhage or brain tumor; we found no evidence of these conditions. However, we did find an Onodi cell with an internal, low-attenuation, expansile bulging mass and punctate calcification, presumed to be an aspergilloma within a mucocele (figure 1). Because of the known close association between Onodi cells and the optic nerve, we re-examined the patient's right eye more closely. She had decreased right visual acuity and right retrobulbar pain. Visual acuity of the right and left eyes was 20/100 and 20/20, respectively. Further ophthalmological examination revealed a central visual field defect in the right eye. The left visual field and eye movements on both sides were normal. To evaluate the optic nerve, we performed brain magnetic resonance imaging (MRI) with enhancement.

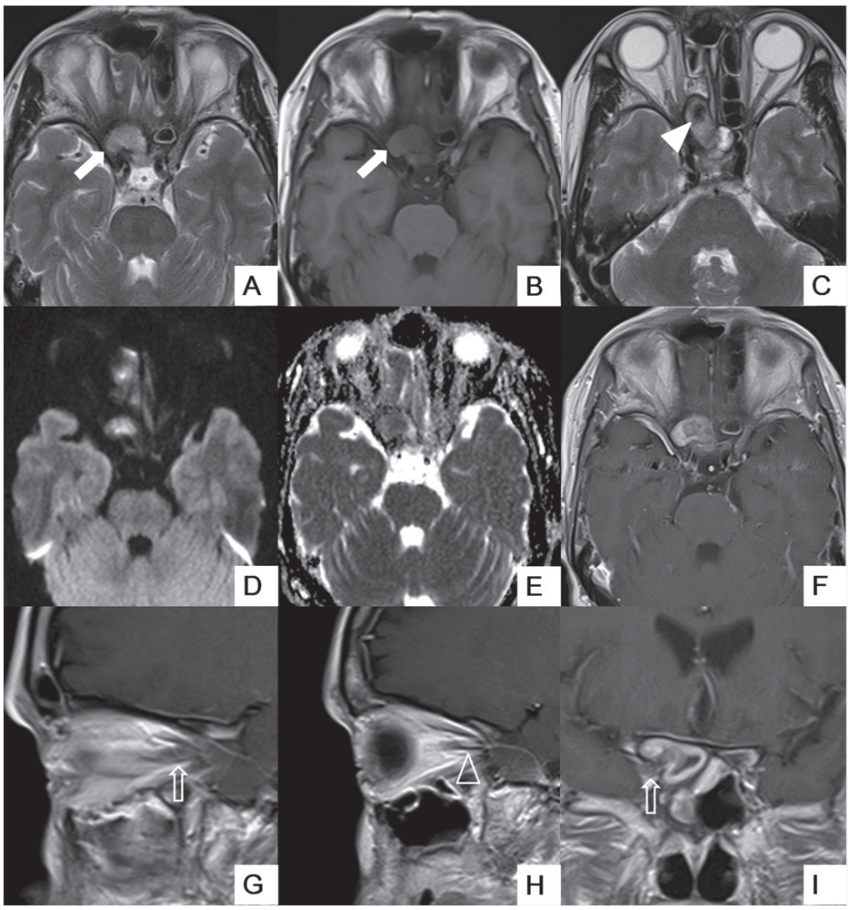

Figure 2.

A-B. Brain magnetic resonance images show cystic, expansile mass (arrows) with iso- to low- signal intensity on axial T2- and T1-weighted images, indicates mucocele.

C. In axial T2-weighted image, a dark signal intensity lesion (arrowhead) within a mucocele is seen, which corresponds to the aspergilloma on brain CT.

D-E. In diffusion-weighted images, mucocele shows diffusion restriction, suggesting an infected mucocele (pyomucocele).

F. In axial T1-weighted contrast-enhanced image, the pyomucocele shows thick rim enhancement

G-H. In sagittal T1-weighted contrast-enhanced images, right optic nerve (blank arrow) is thicker than the left optic nerve (blank arrowhead).

I. In coronal T1-weighted contrast-enhanced image, the pyomucocele shows thick rim enhancement extending over the orbital apex and encasing the right optic nerve. Nerve sheath enhancement of the right optic nerve (blank arrow) is seen, suggesting optic neuritis. However, there is no definite compression of the right optic nerve.

Brain MRI revealed an infected mucocele (pyomucocele), which contained an aspergilloma and extended over the orbital apex, encasing the right optic nerve. Neuritis in the right optic nerve was also seen. However, there was no definite compression of the right optic nerve due to the pyocele in the Onodi cell (figure 2).

The patient was admitted to the Department of Ophthalmology and initially treated with intravenous methylprednisolone (1g per day over a three-day period) according to the optic neuritis treatment protocol. (4) After medical treatment, right visual acuity recovered to 20/25. The patient was then transferred to the Department of Otolarygology for drainage of the pyomucocele. The patient underwent an endoscopic sphenoidectomy and ethmoidectomy; a pyocele containing an aspergilloma was confirmed by histologic examination. The patient experienced dramatic relief of her symptoms and was free from any ocular events during the postoperative follow-up period of 2 months.

DISCUSSION

In 1904, Onodi first described an anatomical variation in which a posterior ethmoidal cell projected within the sphenoid bone. (2) The incidence of Onodi cells is reported to be 8% to 13%; however, an incidence of 33-65.3% was reported in a recent study. (5) Theoretically, various diseases of the paranasal sinuses, including infectious or inflammatory sinusitis, inverted papillomas, aspergillomas, mucoceles, and sinonasal malignancies, may be found in Onodi cells; however, pathologies of Onodi cells are rare. (6)

Patients with Onodi cell infections may present with optic neuritis, and 3 potential etiological mechanisms have been proposed. The first hypothesis is that bulging of the Onodi cell due to infection results in a direct mass effect on the optic nerve, causing venous ischemia and subsequent inflammation. The second hypothesis is that infection in an Onodi cell causes local inflammation and secondary optic neuritis via a cleavage in the optic canal wall or via a bone resorption site. The last hypothesis is that infection in an Onodi cell causes optic neuritis via vascular or lymphatic vessels. (3) In our patient's case, the optic nerve was encased by the Onodi cell, yet definite compression was not seen. Thus, the second or third hypotheses provide more likely explanations for the development of optic neuritis in this case.

Visual field defects due to Onodi cell in-

fections with optic neuritis are common, tend to develop acutely, and are often reversible with prompt diagnosis and timely drainage. (7) However, the visual field defects do not recover if appropriate treatment is carried out more than 6 weeks after the onset of symptoms. (6)

It is important not only to detect Onodi cell infections, but also to differentiate between optic neuritis related to a pyomucocele and compressive optic neuropathy as the treatments for each are somewhat different. Optic neuritis, as in our patient's case, presents as acute loss of vision, headache, and retrobulbar pain, (8) as does compressive optic neuropathy. Contrast-enhanced CT or MRI scans are important for preoperative differential diagnosis and planning of surgery for pyomucoceles and simple mucoceles in Onodi cells. Marked thick rim enhancement in Onodi cells is characteristic of pymucoceles. However, thin enhancement of the mucosal lining is characteristic of simple mucoceles. (9) In optic neuritis, optic nerve swelling, optic nerve sheath dilatation, or optic nerve sheath enhancement can be seen on coronal T1-weighted contrast-enhanced MRI. (10)

Optic nerve neuritis caused by an aspergilloma within a pyomucocele in an Onodi cell is an extremely rare clinical finding. However, as this is a vision-threatening condition, it must be diagnosed and treated promptly by medical and surgical teams to ensure the return of normal visual function. Emergency physicians should consider this diagnosis in patients with headache and decrease in visual acuity, and should conduct imaging studies for accurate diagnosis and to prevent permanent loss of vision.

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