CME Article

Clinics in diagnostic imaging (120)

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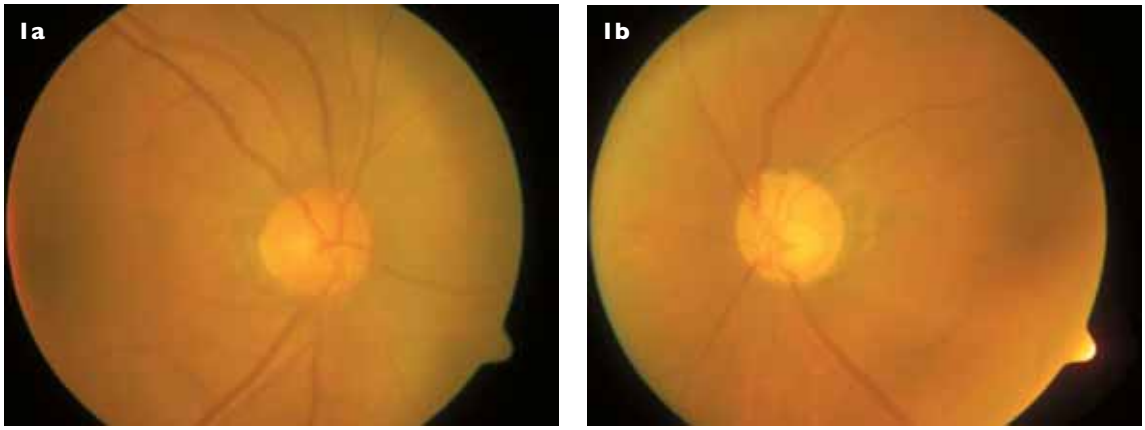

Fig. 1 Colour photographs of the (a) right and (b) left optic nerves.

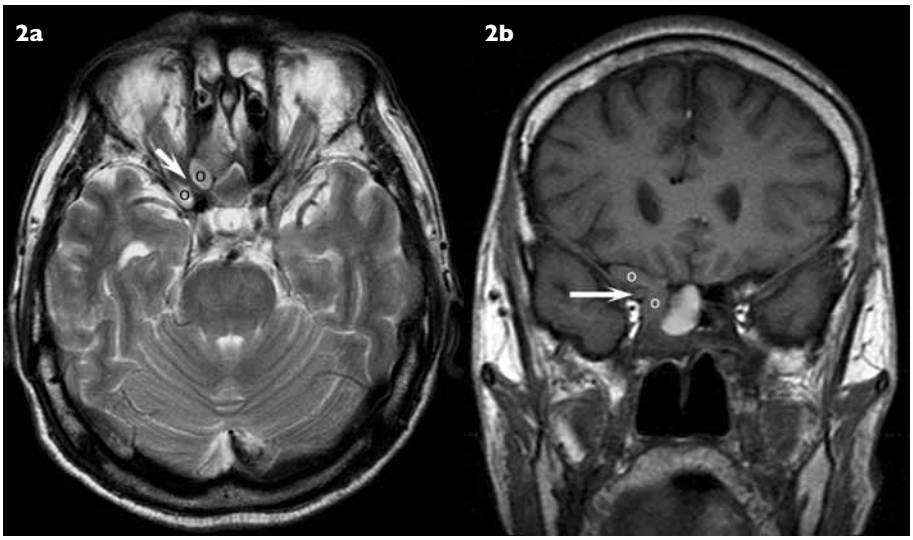

Fig. 2 (a) Axial T2-W and (b) coronal T1-W images of the sinuses.

CASE PRESENTATION

A 60-year-old tailor was working when he noticed a sudden ache on the nasal aspect of his right eye. At the same instance, the vision in his right eye deteriorated and he was unable to differentiate the colours of the garment he was working on. He had a past history of nasopharyngeal carcinoma diagnosed and treated with radiotherapy in 1999, and was presently in remission. In addition, he had an attack of acute angle closure glaucoma affecting his left eye two years ago for which bilateral peripheral iridotomies were performed. He had defaulted further treatment. On examination, the best corrected visual acuity was 6/30 on the affected right eye and 6/12 on the left. Colour vision using the Ishihara plates was decreased on the right (1/15 on the right, 15/15 on the left). Confrontation visual field revealed a temporal defect on the right and a tunnel field on the left. A right relative afferent papillary defect was present. He had bilateral peripheral iridotomies, and intraocular pressures were normal (15 mmHg in each eye).

What do his disc photos show and explain why the affected optic nerve is still pink although he has a relative afferent pupil defect on that side (Figs. 1a–b)? He was provisionally diagnosed with right radiation optic neuropathy. Differential diagnoses were retrobulbar optic neuritis, infective optic neuropathy, compressive optic neuropathy and posterior ischaemic optic neuropathy. Magnetic resonance (MR) imaging of the anterior visual pathway was performed (Figs. 2a–b). What is your diagnosis? What should the subsequent management be?

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IMAGE INTERPRETATION

Colour photographs show that the optic disc was pink on the affected right eye with a cup/disc ratio of 0.3, but was severely cupped on the left. There is cupping of the left optic disc due to the previous acute angle closure glaucoma two years prior to this (Figs. 1a–b). This also explains the tunnel field on that eye on confrontation visual field testing. The pink optic disc on the affected right eye indicates that the pathology is of fairly acute onset and disc pallor, which usually takes 6–8 weeks, has not yet occurred. There is a right relative afferent papillary defect as the right optic nerve is more severely compromised compared to the left due to the current acute problem. MR imaging of the anterior visual pathway revealed no abnormality of the optic nerves (arrowhead shows the right optic nerve) or orbital contents. However, the right posterior ethmoid cells were opacified and extended laterally and superiorly around the right orbital apex, typical of mucocoele involving the Onodi cell (circles), and causing rhinogenic optic neuritis, accounting for the clinical picture (Figs. 2a–b).

DIAGNOSIS

Right rhinogenic optic neuritis secondary to mucocoele of the Onodi cell.

CLINICAL COURSE

The patient was referred to the otolaryngologist, who commenced medical treatment with antibiotics prior to surgical drainage. Computed tomography (CT) of the paranasal sinuses confirmed features of pansinusitis and a right-sided Onodi cell mucocoele (Fig. 3). Functional endoscopic sinus surgery with decompression of the Onodi cell was performed. Intraoperatively, inspissated purulent material was present in both the posterior ethmoid and sphenoid sinuses. The right optic nerve was found to be dehiscent within the Onodi cell (Fig. 4). A few days after commencement of antibiotics and surgery, the vision in his right eye improved and on follow-up, recovered completely (visual acuity 6/6, normal colour vision and resolution of visual field defect). He is presently on follow-up for glaucoma in the left eye. His posterior ethmoid and sphenoid sinuses remain open and clean on endoscopic surveillance at one year postoperation.

DISCUSSION

This case illustrates the rare occurrence of mucocoele formation in the osseous structures forming the optic canal. As such, it is intimately related to the optic nerve. The recognition of Onodi cells is important for two reasons: first, complications of sinusitis may rarely play an important role in the pathogenesis of rhinogenic optic neuropathies, and second, the risk of optic nerve injury, which is a potential and devastating complication of endoscopic sinus surgery, is increased. The prevalence of Onodi cells varies from 7% using CT to 60% by anatomic dissection in cadavers.[1-3] It has been reported to be more prevalent in Asia compared to the Western countries (60% vs. 39%).[1] There is still no consensus on whether CT or MR imaging is superior for demonstrating this abnormality.

The four pairs of paranasal sinuses, named after the skull bones in which they are located, and their involvement by acute or chronic sinusitis, are well known to most physicians. In the case of sinusitis with frontal or maxillary involvement, the common presentations are that of pain and tenderness just over the affected sinus. When the

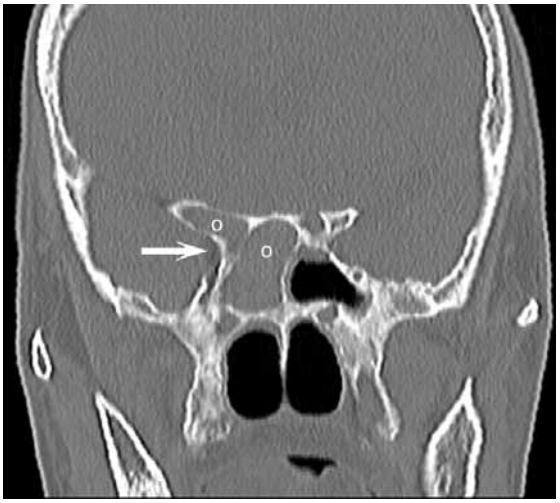

Fig. 3 Coronal CT image of the paranasal sinuses shows sinusitis of the Onodi cell with the right posterior ethmoid cells opacified and extending laterally and superiorly around the right orbital apex (arrow).

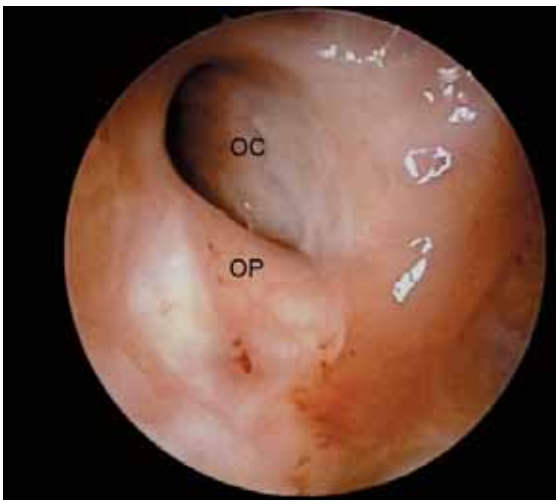

Fig. 4 Endoscopic photograph shows the right Onodi cell (OC). The right optic nerve (OP) was found to be dehiscent within the Onodi cell.

ethmoid or sphenoid sinuses are affected, it can give rise to headache or even extend to the orbit and cause orbital cellulites. However, not many are aware that during the normal embryological development of a sinus, pneumatisation may involve adjacent bones. For example, the maxillary sinuses can extend into the zygomatic bones, and the ethmoid air cells can involve the frontal, sphenoid and maxillary bones.[4] Onodi cell refers to a posterior ethmoidal air cell that has extended into the sphenoid bone.[5] In the third foetal month, anterior and middle ethmoid cells begin as invaginations of the lateral nasal wall in the region of the middle meatus. Shortly after that, posterior cells invaginate the nasal mucosa in the superior meatus and enlarge progressively. In the case of an Onodi cell, the posterior ethmoidal air cells encroach on the adjacent sphenoid bone surrounding the optic canal.[4,5] As a result of its proximity to the optic nerve, sinus infection of Onodi cells can result in contiguous infection of the nerve, and optic neuropathy ensues. Rhinogenic optic neuropathy secondary to complications of sinusitis of Onodi cell, as seen in our patient, is a rare complication.[6-9]

Radiological investigations with CT or MR imaging are vital to the diagnosis of mucocoele or sinus infection of Onodi cells. The MR imaging signal characteristics typical of a mucocoele reflect the protein content of the mucoid material. On T1-weighted sequences, signal intensity is initially decreased, due to its high water content. However, as water is resorbed in the course of time, the protein content and viscosity are increased, resulting in signal intensity that is initially isointense and then hyperintense, relative to the brain on T1-weighted images. On T2-weighted images, the signal intensity usually remains high, but may be decreased as the contents become inspissated.[9] In this case, the heterogeneous nature of the signal on both T1- and T2-weighted images is typical, making the diagnosis of mucocoele of Onodi cell straightforward. This however is not always the case, and when the signal intensity of the mucocoele is isointense with brain on both T1- and T2-weighted images, other differential diagnosis, such as malignant disorders, cannot be easily excluded.[9] The mucous content of sinus mucocoele can also be detected in MR spectroscopy, where N-acetylated glycoprotein may be confused with normal N-acetyl aspartate, which is normally found in the brain.[10]

Mucocoeles of the Onodi cell have been reported to cause unilateral, as well as bilateral, visual loss.[6-8,11] Surgical drainage appears essential for definitive treatment of this condition.[6,11] With the advent of functional endoscopic sinus surgery, this is currently the preferred mode of treatment. This case represents the rare occurrence of mucocoele formation in an Onodi cell resulting in rhinogenic optic neuropathy. A detailed understanding of the anatomy of the paranasal sinuses and its variations is essential not only in making the correct diagnosis, but also in the safe management of the condition. Ophthalmologists, radiologists as well as otolaryngologists should be familiar with the rare occurrence of sinusitis of the Onodi cell causing optic neuropathy. They should also be familiar with the MR imaging signal characteristics of this condition as demonstrated in this case.

ABSTRACT

Acute visual loss can be caused by retrobulbar optic neuritis, radiation optic neuropathy or ischaemic optic neuropathy. Sinusitis affecting the Onodi cell, a posterior ethmoidal air cell that has encroached on the adjacent sphenoid bone forming the optic canal, can present rarely with visual loss. We report a 60-year-old man, who developed a sudden ache on the nasal aspect of his right eye, and deterioration of the vision in his right eye. This case illustrates the typical radiological appearances of the Onodi cell on MR imaging and CT. The diagnosis of right rhinogenic optic neuritis secondary to mucocoele of the Onodi cell was confirmed at surgery. Functional endoscopic sinus surgery with decompression of the Onodi cell was performed. Physicians should be familiar with the presentation, diagnosis and management of this rare but important condition.

Keywords: acute visual loss, Onodi cell mucocoele, retrobulbar optic neuritis, rhinogenic optic neuritis, sphenoethmoidal sinus

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SINGAPORE MEDICAL COUNCIL CATEGORY 3B CME PROGRAMME

Multiple Choice Questions (Code SMJ 200801B)

	True	False
Question 1. Acute visual loss:		
(a) Associated with pain, redness and headache, can be due to acute angle closure glaucoma.	☐	☐
(b) Associated with a swollen optic disc in an elderly man, requires radiological investigations to rule out a mass lesion.	☐	☐
(c) In a young female, associated with pain on eye movements and a swollen disc, is typical of multiple sclerosis.	☐	☐
(d) Associated with an increased cup disc ratio and optic disc cupping, is typical of open angle glaucoma.	☐	☐
Question 2. Onodi cell:		
(a) Refers to a posterior ethmoidal air cell that has extended posteriorly over the anterior wall of the sphenoid sinus.	☐	☐
(b) Is closely related to the optic nerve and requires radiological investigations to detect its presence preoperatively.	☐	☐
(c) Is important, because it requires removal, if present.	☐	☐
(d) Is important, as its presence during endoscopic sinus surgery can lead to inadvertently damaging the optic nerve.	☐	☐
Question 3. Mucocoele of the ethmoid and sphenoid sinuses:	☐	☐
(a) Can be complicated by orbital cellulites and orbital abscesses.	☐	☐
(b) Requires treatment with antibiotics as well as surgical drainage.	☐	☐
(c) Can be diagnosed by symptoms alone.	☐	☐
(d) Can lead to cavernous sinus thrombosis and brain abscesses.	☐	☐
Question 4. In a patient presenting with acute visual loss, signs of optic neuropathy and a normal-looking optic nerve:		
(a) Retrobulbar optic neuritis is a possible cause.	☐	☐
(b) Anterior ischaemic optic neuropathy is a differential diagnosis.	☐	☐
(c) Rhinogenic optic neuropathy is a differential diagnosis.	☐	☐
(d) Radiological investigations should be conducted to rule out compressive lesions.	☐	☐
Question 5. Radiographical findings in patients with mucocoeles:		
(a) Include decreased signal intensity on T1-weighted sequences initially.	☐	☐
(b) Are never hyperintense on both T1- and T2-weighted images.	☐	☐
(c) May mimic malignancies occasionally.	☐	☐
(d) Are not affected by proteins and viscosity.	☐	☐

Doctor's particulars:

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SUBMISSION INSTRUCTIONS:
(1) Log on at the SMJ website: http://www.sma.org.sg/cme/smj and select the appropriate set of questions. (2) Select your answers and provide your name, email address and MCR number. Click on "Submit answers" to submit.

RESULTS:
(1) Answers will be published in the SMJ March 2008 issue. (2) The MCR numbers of successful candidates will be posted online at www.sma.org.sg/cme/smj by 15 March 2008. (3) All online submissions will receive an automatic email acknowlegment. (4) Passing mark is 60%. No mark will be deducted for incorrect answers. (5) The SMJ editorial office will submit the list of successful candidates to the Singapore Medical Council.

Deadline for submission: (January 2008 SMJ 3B CME programme): 12 noon, 25 February 2008.