

A Case of a Mucocele in an Onodi Cell

Manabu Nonaka, Akira Fukumoto, Reiko Nonaka,
Chika Ozu and Shunkichi Baba

Department of Head & Neck and Sensory Organ Science, Graduate School of Medicine, Nippon Medical School

Abstract

Onodi cells have been defined as posterior ethmoid cells that have pneumatized laterally and superiorly to the sphenoid sinus. They are often close to the optic nerve or the internal carotid artery. A patient complained of a sudden decrease in left eye visual acuity and left eye pain due to formation of a primary mucocele in an Onodi cell. A coronal computed tomography scan and magnetic resonance imaging were useful for diagnosing the mucocele. (J Nippon Med Sch 2007; 74: 325–328)

Key words: Onodi cells, primary mucocele, visual disturbance

Introduction

Mucocele lesions of the posterior ethmoid and sphenoid sinuses are rare, but they may be accompanied by visual organ disorders. It is important to start treatment soon after a visual disturbance develops.

The Onodi cell has been defined as a posterior ethmoid cell that has pneumatized laterally and superiorly to the sphenoid sinus and is intimately associated with the optic nerve¹. We report here on a patient in whom a primary mucocele developed in an Onodi cell and caused an acute disturbance.

Case Report

On the morning of November 23, 1998, a 41-year-old Japanese man experienced a left eye pain and a sudden decrease in left visual acuity. The symptoms gradually became worse, so the patient visited a nearby eye clinic on November 25. The right eye

acuity was 1.0 (20/20), whereas that of the left eye was 0.04 (20/500). Left eye neurosis was suspected, and the patient was referred to the department of ophthalmology of Nippon Medical School. At the time of eye examination on November 27, the left eye acuity was limited to light sense. Fundoscopic examination showed redness and swelling of the optic nerve papilla on the left side, and retrobulbar optic neuritis was diagnosed. The patient was also examined in the hospital's department of neurosurgery on the same day. No other neurological abnormalities were found, and a horizontal computed tomography (CT) scan of the head did not reveal any intracranial lesions. Examination of the cerebrospinal fluid did not yield any findings suggestive of demyelinating disease. For 5 days from November 30, the patient was treated with betamethasone sodium phosphate at a dosage of 4 mg/day and hydrocortisone sodium succinate at the dose of 1,000 mg/day, but there was no improvement in the visual acuity. A magnetic resonance (MR) scan performed on December 2

Correspondence to Manabu Nonaka, Department of Otorhinolaryngology, Nippon Medical School Tama Nagayama Hospital, 1-7-1 Nagayama, Tama-city, Tokyo 206-8512, Japan
E-mail: nonaka@nms.ac.jp
Journal Website (<http://www.nms.ac.jp/jnms/>)



Fig. 1 T2-weighted MRI reveals a cystic lesion as a high signal in a 15×15×5 mm region immediately superior to the left optic nerve. (⇔): mucocele; (△): optic nerve.
a: Horizontal section
b: Coronal section

raised the possibility of a paranasal sinus mucocele, and the patient was referred to the authors' department on December 5.

Rhinoscopic examination found no abnormalities. T1-weighted MR images showed a signal similar to that of the substantia grisea in a 15 × 15 × 5-mm region immediately superior to the left optic nerve, and T2-weighted images revealed a high signal intensity cystic lesion (**Fig. 1a, b**). Coronal CT scan of the paranasal sinus showed a soft-tissue shadow superior to the left optic canal. However, there were



Fig. 2 A paranasal sinus coronal-section CT scan shows a soft-tissue shadow (⇔) laterally and superiorly to the sphenoid sinus.

no other shadows in the paranasal sinus (**Fig. 2**).

It was speculated that the loss of visual acuity was due to a paranasal sinus mucocele, and endoscopic mucocele-opening surgery was performed under general anesthesia on December 7. What was thought to be part of a mucocele wall was observed supralateral to the sphenoid sinus at the back end of the posterior ethmoid sinus. A small incision was made there, and discharge of a serous exudate was observed. The incised site was opened as completely as possible. Below the mucocele, the left optic canal prominence was observed. The optic nerve was not exposed.

The left eye pain had resolved on the day after the mucocele surgery, but the visual acuity had not recovered even after 6 months' postoperative follow-up.

Discussion

It is thought that the first reference to the close relationship between the posterior ethmoid sinus and the optic nerve was made by Onodi in 1904². The Onodi cell has been defined as a posterior ethmoid cell that has pneumatized laterally and superiorly to the sphenoid sinus¹. The reported incidence of Onodi cells ranges from 8%³ to 13%⁴ on the basis of CT and radiological findings, and Onodi cells are often close to the optic nerve or the internal carotid artery.

In the case of paranasal sinus mucoceles, the paranasal sinus is filled with exudate, the paranasal sinus is enlarged, and there is displacement and destruction/resorption of the bony wall. As the cause of these changes, narrowing or blockage of the paranasal sinus ostium, inflammation, and other conditions have been proposed⁵, but the cause remains unclear. The mechanism of bone resorption had been thought to involve internal pressure due to retained fluid⁵, but more recently IL-1 and other bone-resorption-promoting cytokines that are produced by the mucocele epithelium have been suggested to be involved⁵. Approximately 90% of primary mucoceles are thought to be formed in the boundary area between the frontal sinus and the anterior ethmoid sinus⁶. Several reasons have been proposed for localization on the basis of anatomical characteristics of the ethmoid sinus, including the fact that the ethmoidal cells have an overall shape of an arrowhead, so that they become progressively narrower in the forward direction, that the cells of the anterior ethmoid sinus are more numerous than those of the posterior ethmoid sinus, and that the ostia of the frontal sinus and the anterior ethmoid sinus are narrow⁷. In contrast, the ostia of the posterior ethmoid sinus and the sphenoid sinus are wide, resulting in a low incidence of mucocele formation. In particular, formation of a primary mucocele in an Onodi cell, as seen in our patient, is extremely rare, and our search of the literature found reports of only two earlier similar cases^{2,8}.

Two causative mechanisms for the visual disturbances have been proposed⁹. According to one theory, cleavage of the optic canal wall or bone resorption occurs, the optic nerve is directly compressed by the mucocele, and ischemia of the nerve and venous congestion develop. In particular, it is thought that because the inside of the optic canal is not lined with fat or other soft tissues, pressure that is applied externally is readily transferred directly to the nerve. The second theory postulates that inflammation occurs due to an infection in the mucocele and spreads to the nerve via a cleavage in the optic canal wall or via a bone resorption site. In our patient, gross observation performed during the operation did not reveal the

optic nerve to be exposed, and for this reason it can be surmised that inflammation had spread via a cleavage in the optic canal wall or a small site of bone resorption caused by the mucocele. The visual disturbance caused by a mucocele can sometimes be temporarily reversed by administration of a corticosteroid, but the best therapeutic approach is early surgery to open the mucocele⁹. In general, the prognosis is poor when the preoperative degree of visual impairment is high or when its onset was very sudden. In cases with a mild to moderate degree of visual impairment and gradual progression, the prognosis depends on the time from onset until surgery is performed; the prognosis is poor when 1 or 2 months have passed since onset⁹. It has also been reported that even when the degree of visual impairment is high, recovery of visual acuity is achieved when surgery is performed within 2 days after the decrease in visual acuity occurs and within 24 hours of the loss of light sense¹⁰. Thus, it is desirable for surgery to be performed quickly. In our patient, a severe visual impairment was observed on the fourth day after onset of the left eye symptoms. Fourteen days had passed from onset until surgery, and recovery of the patient's visual acuity has not been observed.

One reason that the discovery and diagnosis of the mucocele was late was that the horizontal CT scan obtained by the department of neurosurgery did not reveal the mucocele. Ogata et al.⁸ discussed two problems associated with lesions of an Onodi cell. One is that it is easy for an Onodi cell to escape detection when only a horizontal-section CT scan is performed. The posterior ethmoidal cells are 8 to 20 mm in size, and there is a possibility that an Onodi cell itself would not be detected when using a slice width of 10 mm or more. In addition, Onodi cells are generally flat and are positioned laterally and superiorly to the sphenoid sinus. Thus, Ogata et al. pointed out that a coronal-section CT scan and MR are useful for the diagnosis and evaluation of Onodi cells. The second problem raised by Ogata et al. is that we lack anatomical knowledge regarding Onodi cells. In our patient a coronal-section CT scan and MR were useful for the diagnosis. The horizontal-section CT alone was unable to reveal the lesion in

the paranasal sinus, and there is a possibility that it would have escaped detection in the absence of anatomical knowledge regarding the existence of Onodi cells and knowledge that a mucocele limited to that area could develop and if a coronal-section CT scan had not been performed.

Surgery can be expected to lead to improvement in the visual impairment caused by a paranasal sinus mucocele, but the prognosis is influenced by the time interval from onset to surgery. In particular, when the visual impairment is severe, it is desirable that surgery be performed within 24 hours of onset. As in our patient, visual impairment can occur even when the mucocele lesion is small and restricted to an Onodi cell. Accordingly, we suggest that MR and a coronal-section CT scan are necessary for the evaluation of retrobulbar optic neuritis.

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