CASE REPORT

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Complicated Onodi cell mucocele presenting with orbital apex syndrome; a case report and literature review

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Abstract

Mucocele is defined as the accumulation of mucus within a cystic structure lined by mucoperiosteum causing bone remodeling and erosion. The sphenoidal sinus mucoceles, due to its proximity to the optic nerve, can cause ocular complications. A50-year-old woman with a history of previous endoscopic nasal surgery (30 months ago), blurred vision, severe right-sided orbital pain since three weeks ago and severe unilateral headache was admitted. Mucocele related orbital apex syndrome due to the sphenoid sinus mucocele was diagnosed. The optic nerve was fully dehiscent and under compression. Paranasal sinus pathologies must be taken into consideration when a patient with the diagnosis of optic neuropathy and involvement of the superior orbital fissure is approached.

INTRODUCTION:

The paranasal sinuses are prone to ostium obstruction due to surgery, inflammation, trauma and tumors (1), which can result in mucocele formation (2). Mucocele is defined as the accumulation of mucus within a cystic structure lined by mucoperiosteum causing bone remodeling and erosion (3). The paranasal sinuses' mucocele occurs in 30 to 60 years old adults. Frontal mucocele accounts for 65% of such cases, while ethmoid and maxillary sinus mucoceles have a prevalence of 25% and 10%, respectively. Rare mucocele types consist of sphenoid and Onodi cell account for less than 1% of paranasal mucoceles (1,4,5). In 1904, Onodi cell was described for the first time as the anatomic variant of posterior ethmoidal cells projected into the sphenoid. Today Onodi stands for the most posterior ethmoidal cells which have been pneumatized superior to the sphenoid. It can be detected on the CT scan in 7% of the general population, but could be discovered in a cadaver in up to 60% of the cases. This difference is most probably due to the lack of sufficient resolution of CT scan images (4,6). Although it has been reported that the sphenoid and posterior ethmoid mucoceles are not rare in Japan, yet around 78% of such cases are post-up patients (5). Mucocele is generally formed 5.3 years after performing FESS (Functional Endoscopic Sinus Surgery), 17 years after maxillofacial trauma without any history of surgery and 18.1 years after open sinus surgeries (3). Mucoceles are capable of expansion due to the production of osteolytic factors such as

prostoglandines and collagenases. The incidence of intraorbital and intracranial complications are around 35 to 47% (3,7). Due to the proximity of Onodi cell mucoceles to the optic nerve, they can cause ocular complications (2).

CASE REPORT

A50-year-old woman with blurred vision, severe right-sided orbital pain since three weeks ago and severe unilateral headache was admitted to our hospital. Her vision at presentation (on admission) was counting fingers (CF) at 30 cm in her right eye (OD), while the left eye (OS) vision was 9/10. Relative afferent pupillary defect (RAPD, Marcus Gunn pupil) was present in OD upon presentation with corresponding mild optic disc pallor on fundoscopy. Visual field could not be assessed in the right eye (due to the patient's poor cooperation). She reported severe pain when moving eyes (ophthalmodynia). The right eye was exotropic and hypertopic in the primary position with limited movement in all gaze directions (ophthalmolplegia). Ptosis was not present on the right side; however, mild proptosis was identified. Diplopia was not determined probably due to the poor vision and also suppression of the right eye images. OS was completely normal on examination. She had no history of asthma or smoking. However, her medical history revealed hypertension and allergic rhinitis. The patient had undergone endoscopic sinus surgery 30 months ago with the possible diagnosis of nasal polyposis, but the pathology examination result was not available. Moreover, she had been admitted to the neurology ward 18 months before with the complaint of blurred vision in her right eye. She was treated medically resulting in a rapid improvement (type of administered medications are not known). The patient was this time admitted to the neurology ward with the complaints of sudden blurred vision and ophthalmoplegia. A consultation with the ENT surgeon was requested, and high septal deviation to the left and nasal polyps medial and lateral to the middle turbinates, without any evidence of Mucormycosis or necrosis was reported in the nasal endoscopy. The sphenoid sinus orifice had a polypoid pattern. The opacification and expansion of sphenoid and right posterior ethmoidal sinuses were diagnosed in CT scan examination. Moreover, a hypodense lesion with erosions in the superior and lateral sphenoidal walls and the posterior part of the lamina papyracea with expansion into the orbital apex, causing a compressive effect on the optic nerve was observed. The optic canal was dehiscent; No bone ridge was seen in the lateral wall of the mucocele, probably due do the previous surgery or mucocele-related erosion. The right maxilla and frontal sinuses were also opacified (Fig. 1, A). In Magnetic Resonance Imaging(MRI) evaluation, a cystic mass with expansion towards the optic nerve was detected in the orbital apex, which was hypointense in T1 and had no enhancement with contrast material. In T2, it was hyperintense as expected (Fig. 1, B). Given the involvement of all the 2nd, 3rd, 4th and 6th cranial nerves, mucocele related orbital apex syndrome was diagnosed. The patient underwent FESS with the guide of the imaging navigation system. The right middle turbinate was shaved and antrostomy, anterior and posterior ethmoidectomy and right sphenoidotomy were performed. Under the navigation guide, the suspicious lesion was approached and a considerable amount of purulent material came out. The optic nerve was fully dehiscent and exposed while the mucocele had a compressive effect on it. At this point we realized that the mucocele is placed in the Onodi region (Fig. 1, C); therefore, the medial wall of the mucocele was removed. The treatment with intravenous antibiotics and corticosteroids was continued postoperatively. On the first post-operative day the right eye movements returned to normal, but the vision was still CF at 40-50 cm, which improved to 7-8/10 during the next three weeks.

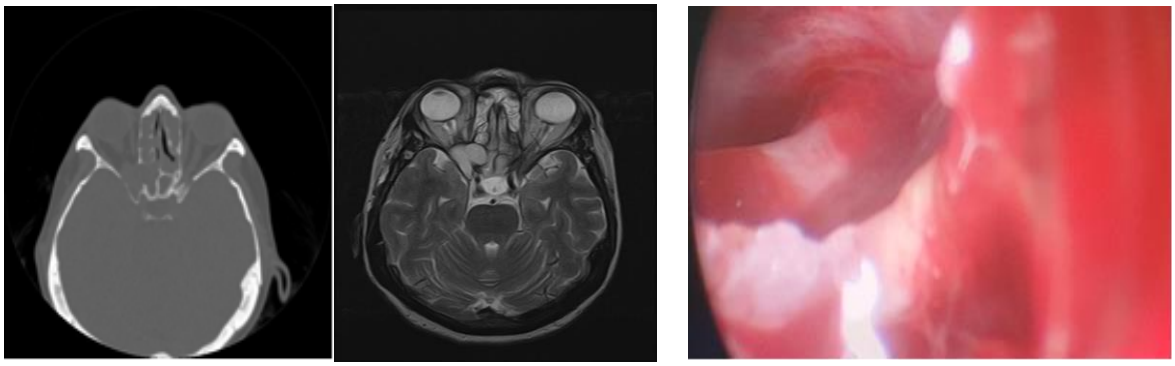

Figure 1. (A) Computed Tomography, (B) Magnetic Resonance Imaging and (C) Intra-operative Endoscopic picture of the optic nerve and Onodi cell mucocele.

DISCUSSION

Onodi cell mucoceles are particularly prone to ocular complications due to their proximity to the optic nerve. There is a higher risk of optic nerve dehiscence in the presence of these sphenoethmoidal cells. The optic nerve may also be exposed in the superior lateral wall of the Onodi region. Therefore, the diagnosis of Onodi cell prior to surgery is of major importanc (2,6,8). Sphenoid or sphenoethmoid mucoceles can rarely result in orbital apex syndrome, retobulbar neuritis or imitate sellar or parasellar masses (5). The mechanisms of retrobulbar optic neuropathy caused by mucocele are as follows: 1. Direct mechanical compression of the optic nerve which means gradual increase in the pressure results in erosion of the adjacent bone in the areas with the least resistance; 2. Secondary circulatory disturbance and vascular thrombosis are caused by the compression resulting in ischemic changes; and 3. The inflammatory reaction associated with the mucocele. In the last two cases, ocular symptoms progress rapidly. The sudden onset and rapid progression of visual symptoms show that the mechanism is associated with optic neuropathy and not mechanical compression (8). Moreover, sphenoethmoidal mucoceles can cause sudden or gradual Unilateral visual loss which is usually accompanied by pain (6). Infected sphenoethmoidal cells without a mucocele can also lead to optic neuropathy and orbital apex syndrome (6). Factors indicating poor prognosis are sudden onset of symptoms and poor visual acuity at the time of manifestation (9). Several studies have reported that ethmoidal mucoceles can result in optic neuropathy; however, ethmoidal mucocele causing orbital apex syndrome has been rarely reported (7). The main clinical manifestation of orbital apex syndrome is optic neuropathy accompanied by ophthalmoplegia (10). The term "orbital apex syndrome" is usually used when all structures in the orbital apex and superior orbital fissure are affected due to a disease course, such as the involvement of the 2nd, 3rd, 4^{th}, 6^{th} and the first branch of 5^{th} cranial nerves. The patient may also experience ptosis, proptosis, diplopia, ophthalmodynia, ophthalmoplegia, eyelid edema and visual loss (7). Optical apex syndrome with total ophthalmoplegia is much rarer than the partial type (5). The main etiologies for orbital apex syndrome include: inflammation consisting of idiopathic orbital inflammation and collagen vascular disease; infections such as aspergillosis or

mucormycosis; neoplasia such as nasopharyngeal carcinoma/neurinoma/lymphoma, metastasis, vascular causes, trauma such as sinus surgery and others (mucocele). In Erapan's study, the most common etiology for orbital apex syndrome was carotid cavernous fistula and neoplasms, most commonly lymphomas (10). The diagnosis of mucocele is mainly based on imaging (7). An expanded sinus with bone erosion and wall thinning is seen on CT scan evaluation. Differentiation from an underlying tumor is based on MR imaging (4). Furthermore, MRI is used for determining the lesion's expansion and protein material. In MRI the mucoceles appearance is associated with protein concentration and its mucoid secretion content. It may be hypo, iso or hyper intense or it may be even signal void. T1 Low intensity signal and T2 high intensity signal can typically differentiate a mucocele from other masses (7). Most mucoceles are non-infectious and manifest gradually. The rapid progression of symptoms is seen in infectious mucoceles (mucopyoceles). A considerable number of mucoceles are bacteria-free on laboratory cultures, but as we are unaware of which mucocele is infectious, prior to surgery, antibiotic therapy is administered (1). The standard treatment of mucocele-related orbital apex syndrome is early surgical decompression and drainage with appropriate post-op intravenous antibiotic therapy (1, 5). Endoscopic sinus surgery must be considered regardless of the disease duration. No obvious "visual loss duration" is predictive of the visual outcome. However, the prognosis might deteriorate one to two months after symptoms' onset. In another study it was observed that optic neuropathy recovers if the surgery is delayed even more than 10 days (11). Other factors such as the acute development of symptoms may also affect the prognosis (2). The visual recovery probability is strongly related to the visual acuity status before surgery. If no light perception existed before surgery or is accompanied by sudden visual loss, the prognosis is usually poor (12).

CONCLUSION

In conclusion, the paranasal sinus pathologies must be taken into consideration when a patient with the diagnosis of optic neuropathy and involvement of the superior orbital fissure is approached.

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