Mucocele of the Sphenoid Sinus as a Rare Cause of Isolated Oculomotor Nerve Palsy: Case Report and Review of the Literature

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Abstract

Mucocele of sphenoid sinus is a rare cause of oculomotor nerve palsy. A 59-year-old male presented with a 2-week history of sudden ptosis of his right eyelid and right internal ophthalmoplegia. Laboratory examination and radiological evaluation revealed mucocele of the right sphenoid sinus with diabetes and chronic rhinosinusitis. The patients underwent nasal endoscopic surgery and the right sphenoid sinus mucocele was confirmed. The patient recovered completely after 2 weeks. One year follow up indicated no recurrence. A sphenoid sinus mucocele presents various clinical symptoms due to its relationship to the adjacent cavernous sinus and the base of skull. Involvement of isolated oculomotor nerve palsy in sphenoid sinus mucocele is rare. The differential diagnosis between compressive lesion caused by mucocele of sphenoid sinus and ischemic lesion caused by diabetes should be noticed. Careful clinical evaluation and appropriate radiologic imaging can help to identify a definitive cause. Computed tomography and magnetic resonance imaging are useful in explaining the clinical symptoms of mucocele of sphenoid sinus by identifying the spreading of the lesion. Proper and timely endoscopic marsupialization of the mucocele lead to complete resolution of the oculomotor nerve palsy.

Keywords: Mucocele; Sphenoid sinus; Oculomotor nerve palsy; Ptosis

Introduction

Mucoceles are benign, encapsulated, expansile, locally invasive masses within a paranasal sinus filled with mucus and lined by epithelium. Sphenoid sinus mucocele is the most rarely afflicted sinus and comprise 1-2% of all paranasal sinus mucoceles [1]. Mucocele of the sphenoid is usually seen in fourth decade of life. Once the mucocele expand outside the bony wall, various vital complications may occur since several important structures are adjacent to it. Due to its concealing symptoms, patients usually consult ophthalmologist and neurologist first and easily got wrong or delayed treatment.

Case Presentation

A 59-year-old male, presented with a 2-week history of sudden complete drooping of his right eyelid (Figure 1). His visual acuity was normal with no obviously diplopia, ptosis, facial numbness nor headache. The patients firstly sought medical advice in neurology clinics. Examination revealed uncontrolled hypertension and diabetes. He was initially considered as diabetes-related third nerve palsy and given insulin therapy. However, the symptoms had no obvious relief. Cranium computed tomography (CT) scan was then performed and images revealed soft tissue masses in nasal sinuses. He was then referred to our clinic. Past history showed the patients had nasal surgery 10 years ago and didn’t complain about obvious nasal obstruction and rhinorrhea.

Investigation

Physical examination revealed severe reduction of elevation, depression, and adduction in the right eye. He had a dilated and unresponsive right pupil. The fundus examination and other eyes was unremarkable. Nasal endoscopy showed multiple polyps derived from middle turbinate and occupied nearly the whole nasal cavities accompanied by purulent secretion. The olfactory fissure was also full of nasal polyps and ostium of sphenoid sinus could not be detected bilaterally (Figure 2). Cultures of nasal purulent secretion showed no growth of specific bacteria and fungus.

CT imaging of the nose and paranasal sinus revealed extensive soft tissue masses filled in all sinuses bilaterally and an obvious expansion of right sphenoid sinus. Discontinuity of posterolateral

bony wall of the right sphenoid sinus was remarkable, while the other part of bony wall was thickened. Magnetic resonance image (MRI) was used to evaluate the extension of the lesion. The lesion of right sphenoid sinus showed iso-intense in T1-weighted images and hyper-intense in T2-weighted images with a clear ring shape margin and expand posterolaterally into the right cavernous sinus. The imaging features suggested that the lesion was predominantly cystic (Figure 3). Normal size and shape of pituitary gland, normal optic-chiasma and nerve complex, homogenous enhancement of cavernous were all detected. No evidence of any aneurysm in the circle of Willis in MRA.

Treatment and outcome

The patient underwent functional endoscopic sinus surgery bilaterally and drainage of the right sphenoid sinus under general anesthesia. Intraoperatively, we expanded and removed the anterior wall of right sphenoid sinus in order to allow adequate drainage into sphenethomoidal recess. The secretion of the right sphenoid sinus was serous and purulent. The posterolateral wall bone was discontinuity but the mucus of sinus remained completed. His ptosis was markedly reduced the following day. The patient’s symptom completely diminished 2 weeks after surgery. The patient has been followed in our outpatient clinic at regular intervals. On follow-up
nasal endoscopy there was a healthy mucous membrane lined cavity widely open with no signs of recurrence nor other complication have been found. The patient remains asymptomatic up to now (Figure 4).

**Discussion**

In adults the most common cause of acute oculomotor mononeuropathies is diabetes-related microvascular ischemic third nerve palsy, which is characteristic of various internal ophthalmoplegia with papillary sparing. Papillary sparing is believed to result from microvascular ischemia of the central portion of the nerve with sparing of the more peripherally placed parasympathetic fibers in other words, the iris sphincter is usually unaffected by ischemic injury [2]. In contrast, compressive lesions such as aneurysms or tumors that produce third nerve palsy commonly presenting ocular motility disturbance and iridoplegia at the same time. Sphenoid mucocele is one of rare compressive causes of oculomotor nerve palsy, which occur rarely and has an incidence of 1% of paranasal sinus mucocele [3]. The importance of sphenoid sinus mucocele presenting with isolated third nerve palsy lies in the differential diagnosis. However the pupil-sparing cannot help to precisely distinguish these two kinds of etiologies. The past reported frequency of pupil involvement in several series that included up to 25 patients with diabetes-associated oculomotor nerve palsy ranges from 14% to 32% [4]. What’s more, other reports showed that the sphenoid sinus lesion could also cause isolated pupil-sparing oculomotor nerve palsy, indicated that mucocele probably cause symptoms by compressing the microvascular supply to the nerve, resulting in ischaemia [5,6]. Thus, careful clinical evaluation of identify a definitive cause is important for differential diagnosis between compressive lesion caused by mucocele of sphenoid sinus and ischemic lesion caused by diabetes, especially for ophthalmologist and neurologist.

Paranasal sinus mucocele is commonly defined as the accumulation and retention within a sinus of its mucoid secretion and erosion of one or several of its bony walls. The mechanism in the development of mucocele is not clear, but postulated as the obstruction of the sinus initiates its development [1]. The secretions accumulated within the sinus have no exit and a gradual expansion of the cavity occurs due to the release of osteolytic prostaglandins and mediators with bony remodeling and erosion [7]. In our case, the ostium of right sphenoid is stenosis approached to close completely, possibly caused by potential congenital abnormality. What’s more, the patient suffered from chronic rhinosinusitis for decades and didn’t receive the regular and adequate therapy. Thus the inflammation of sinus, especially the right posterior group of nasal sinuses, would enhance the obstruction of the right sphenoid sinus. Above all, these would be the potential precipitating factors of sphenoid mucocele of this patient.

The mucocele of sphenoid sinus present no symptoms in the early stage. In the late stage, it usually has varied presentations mostly related to the direction of extension towards neighboring structures. Headache is the most common symptom in 70-80% patients, probably caused by involving and irrigating the dura [1]. Expanding sphenoid sinus mucocele may compress the optic nerve or the cavernous sinus which carries the III, IV, V and VI nerves (oculomotor, trochlear, trigeminal and abducent nerves), causing visual disturbance or III, IV, V and VI nerves palsy [8-10]. Oculomotor nerve involvement was reported to account for 70% of ocular palsies [6]. The oculomotor nerve is affected more frequently than the trochlear and abducent nerve but mostly accompanied with palsy of optic nerve [11]. The sole oculomotor palsy caused by sphenoid mucocele is very rare. In our case, it should noticed that the bony wall of right sphenoid sinus is entirety thickening, especially the anterior bony wall. Thus the sphenoid mucocele extended forward to the lateral and posterior direction and invasion into cavernous sinus, which in the end compress the oculomotor nerve and cause symptoms. Table 1 displayed the characteristic of past reports of sphenoid sinus mucocele presenting as third nerve palsy. The patients were mostly mid-aged and ptosis was the most frequent symptom.

A large mucocele produces a classic radiographic appearance of an enlarged distorted sinus with a large bony defect representing a breakthrough into the adjacent structures [6]. CT scan of nasal and paranasal sinuseswould shows a hypo-dense cystic lesion in the sphenoid sinus and bony erosion. In our case, CT showed extensive soft tissue masses in all sinus and bony discontinuity of sphenoid wall, which also indicated fungus rhinosinusitis and inverted papilloma. The contrast medium would help to differential diagnosis. There is usually no increase in the intrinsic density of mucocele after injection of contrast medium due to its avascular mucoid content [6]. The features can be hypo-, iso-, or hyperintense or signal void on the MRI images, depending on their stage of development and protein content [2]. Our case have moderate signal intensity on T1 weighted images and a high signal intensity on T2 weighted images, with peripheral enhancement after administration of contrast, indicating comparatively water-ish content which confirmed during the surgery. What’s more, it is also necessary to exclude potential haemorrhage and cerebral aneurysm by using MRI and MRA, which are the most common and fatal causes for isolated oculomotor nerve palsy. Imaging is important in early diagnosis and help identifying the characteristic of the lesion. CT and MR imaging can supply a reliable diagnosis of mucocele of sphenoidal sinus and give guidance to surgery.

Treatment of sphenoid mucocele is surgical. Traditionally, the management of sphenoid mucoceles was complete removal via transfacial or transcranial approach. However, transnasal sphenoidectomy has largely replaced the conventional open method with excellent results. The endoscopic transnasal sphenoidectomy with sufficient removal of the anterior and inferior walls of the sinus is now strongly recommended to allow adequate drainage of the sphenoid sinus and to avoid recurrences [12]. The prognosis of oculomotor

<table>
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<tr>
<th>Author</th>
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<tr>
<td>Lee et al. [13]</td>
<td>Male, 49</td>
<td>Ptosis, diplopia, headache</td>
<td>Normal</td>
<td>1w</td>
</tr>
<tr>
<td>Akan et al. [6]</td>
<td>Female, 64</td>
<td>Ptosis, medial-gaze paralysis</td>
<td>Null</td>
<td>4w</td>
</tr>
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<td>Vaphiades and Roberson [2]</td>
<td>Female, 90</td>
<td>Binocular diplopia</td>
<td>Normal</td>
<td>4w</td>
</tr>
<tr>
<td>Kataria et al. [14]</td>
<td>Male, 60</td>
<td>Headache</td>
<td>Normal</td>
<td>4w</td>
</tr>
<tr>
<td>Mohbessi et al. [5]</td>
<td>Female, 37</td>
<td>Ptosis</td>
<td>Null</td>
<td>4w</td>
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</table>

Table 1: The characteristics of the past reports of sphenoid sinus mucocele presenting as the oculomotor nerve palsy.
nerve palsy is comparatively better than the optic nerve. The average recover time is 1-4 weeks (Table 1). Proper and timely endoscopic marsupialization of the mucocele lead to complete resolution of the oculomotor nerve palsy.

**Conclusion**

We reported a rare case of mucocele of sphenoid sinus as a cause of isolated oculomotor nerve palsy. The differential diagnosis between mucocele of sphenoid sinus and diabetes should be noticed by clinic surgeon. Careful clinical evaluation and appropriate radiologic imaging can help to identify a definitive cause and give guidance for surgery. CT and MRI are useful to identify the extension of the lesion. Endoscopic marsupialization of the mucocele lead to complete resolution of the oculomotor nerve palsy and needed to be performed properly and timely.

**References**