

Case Report

Orbital apex syndrome caused by ethmoid sinus mucocele: a case report and review of literature

Li-Bo Dai^{1*}, Chao Cheng^{2*}, Jiang Bian², He-Ming Han¹, Li-Fang Shen¹, Shui-Hong Zhou¹, Yang-Yang Bao¹, Jiang-Tao Zhong¹, Er Yu¹

¹Department of Otolaryngology, The First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou 310003, Zhejiang Province, China; ²Department of Otolaryngology, People's Hospital of Jinhua City, Jinhua 321000, Zhejiang Province, China. *Equal contributors.

Received October 15, 2016; Accepted November 16, 2016; Epub January 15, 2017; Published January 30, 2017

Abstract: Ethmoid sinus mucoceles are benign, expansile and cyst-like lesions, when sufficiently large, may causing compression of the optic nerve and nearby structures. We report an extremely rare case of ethmoid sinus mucocele causing orbital apex syndrome. A 59-year-old female presented with over one month history of left-side headache that worsened with left-side ophthalmodynia for six days, accompanied by left-side sudden ptosis and vision loss for half a day. Clinical findings were proved with that of a combined CN II, III, IV and VI paralysis. Computed tomographic scan demonstrated a dense homogeneous mass expanding the left ethmoid sinus and rarefaction of the lateral wall of the left ethmoid sinus with the contents compressing the optic nerve. She underwent a prompt endoscopic sinus surgery. Three days after the operation, the movement and vision of the left eye returned to normal, the left eye pain and headache had also resolved.

Keywords: Ethmoid sinus mucocele, orbital apex syndrome, endoscopic sinus surgery

Introduction

Paranasal sinus mucoceles arise most commonly in the frontal sinus followed by the ethmoid, maxillary and sphenoid sinuses [1]. The clinical manifestation of ethmoid sinus mucocele is dependent on the degree of the mass involvement which includes cranial nerves II through VI [2]. However, ethmoid sinus mucocele causing orbital apex syndrome is extremely rare. Therefore, although benign, when sufficiently large, ethmoid sinus mucoceles are sight-threatening and can lead to permanent visual loss [3]. It should be diagnosed and treated immediately.

Here we report a rare case of orbital apex syndrome caused by an ethmoid sinus mucocele, which was resolved after removal of the mucocele by an endoscopic sinus surgery, along with a review of the relevant English-language literature. This is the first case, to the best of our knowledge, of full recovery of visual function and eye movement in a patient with orbital

apex syndrome secondary to ethmoid sinus mucocele.

Case report

A 59-year-old female presented with over one month history of left-side headache that worsened with left-side ophthalmodynia for six days, accompanied by left-side sudden ptosis and vision loss for half a day. She denied nasal obstruction, postnasal drainage, hyposmia, epistaxis, facial numbness and any impairment in her right visual acuity. She was a non-smoker, non-drinker and had no history of nasal or paranasal sinus inflammation or surgical manipulation.

On ophthalmic examination, there were red and swollen of the left eyelid, ophthalmoptosis and only light perception of the left eye visual acuity. Extra-ocular motility was limited in all directions and a left relative afferent pupillary defect was detected. Ophthalmic and neurological examination of the right eye was normal. On otolaryngological examination, no positive finding was

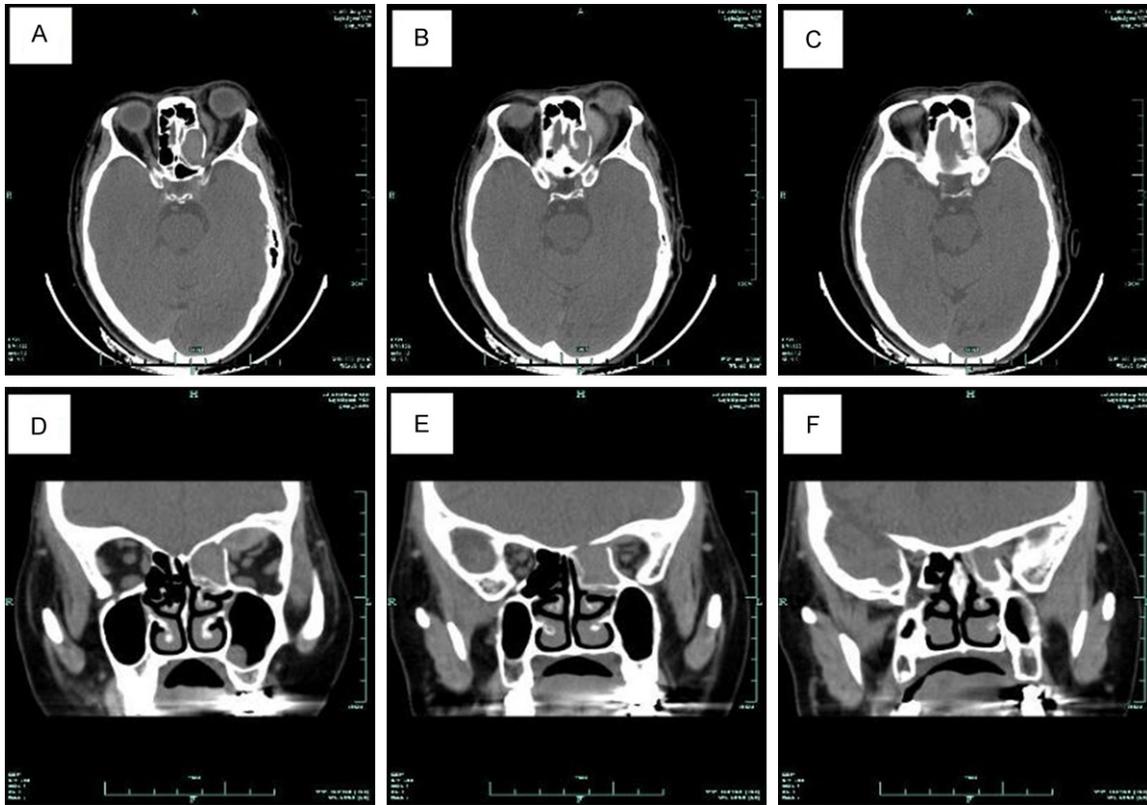


Figure 1. Axial (A-C) and coronal (D-F) CT scan demonstrated a dense homogeneous mass expanding the left ethmoid sinus and rarefaction of the lateral wall of the left ethmoid sinus with the contents compressing the optic nerve.

observed. She was a febrile with no sign of infection.

Clinical findings were proved with that of a combined CN II, III, IV and VI paralysis; therefore the differential diagnoses of orbital apex syndrome was considered and urgent computed tomographic (CT) was performed. CT scan of the paranasal sinuses demonstrated a dense homogeneous mass expanding the left ethmoid sinus and rarefaction of the lateral wall of the left ethmoid sinus with the contents compressing the optic nerve (**Figure 1**).

On the basis of these findings, the patient was diagnosed as orbital apex syndrome secondary to ethmoid sinus mucocele and immediate evacuation and drainage of the cyst was performed by endonasal endoscopic procedure under general anesthesia. On operation, resection of the septum revealed a large ethmoid sinus mucocele containing thick viscid yellow material. Histopathology revealed that respiratory epithelium and inflammatory infiltrate lin-

ing the cyst wall, fibrosis and abundant myxoid stroma were also found, consistent with the diagnosis of mucocele (**Figure 2**). The patient was commenced on a three-day course of 80 mg intravenous methylprednisolone and a week course of cefuroxime. Three days after the operation, the movement and vision of the left eye returned to normal, the left eye pain and headache had also resolved.

Discussion

Mucoceles are epithelium-lined mucus-containing mucoid secretions completely filling a paranasal sinus due to the obstruction of the sinus ostium [4]. The obstruction can be due to infection, allergy, chronic inflammation, congenital anomalies, trauma, iatrogenic injury and tumour [2, 3]. The most common site of occurrence of paranasal sinus mucocele is the maxillary sinus (50%), followed by frontoethmoidal (31%), ethmoidal (16%) and sphenoidal (3%) [3]. Mucoceles are capable of expansion, aided by the production of osteolytic factors,

Orbital apex syndrome due to ethmoid mucocele

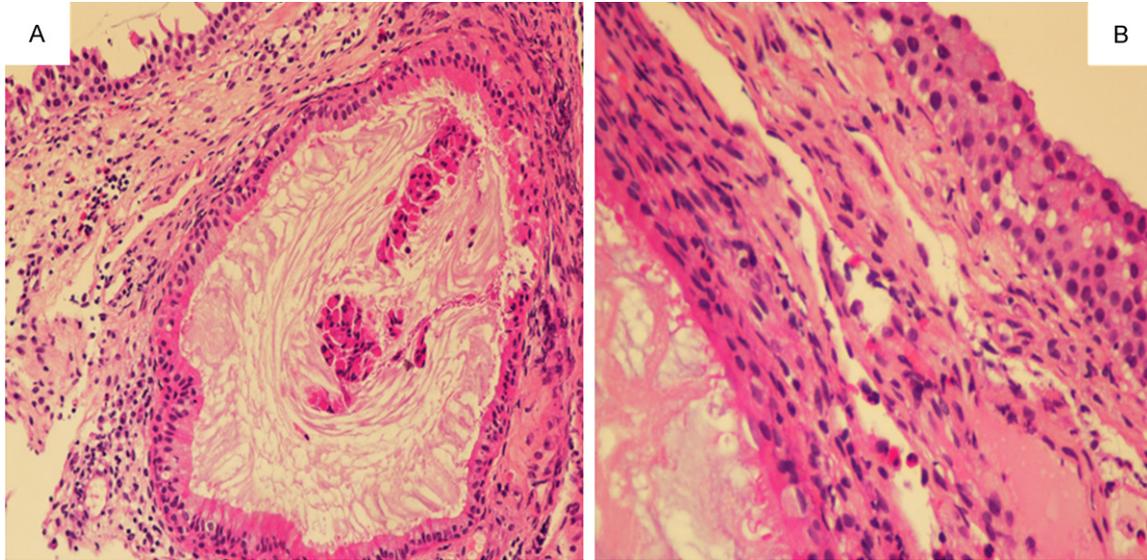


Figure 2. Pathological results showed respiratory epithelium and inflammatory infiltrate lining the cyst wall. Fibrosis and abundant myxoid stroma were present. Hematoxylin-eosin stain, $\times 200$ (A), $\times 400$ (B).

Table 1. Clinical features and final visual outcome of 4 patients with paranasal mucocoeles causing orbital apex syndrome in the English-language literature

Ref	Sex/age	Sinus involved	Symptoms	Visual outcome
Present case	59/F	L ethmoidal sinus	Headache L ophthalmodynia L eyelid swelling L ptosis and vision loss	Completely recovered
Fleissig [11] 2014	53/F	R ethmoidal sinus (Onodi cell)	R vision loss R eye pain R eyelid swelling	NLP
Kumagai [2] 2003	61/M	R sphenothmoid	R proptosis completely R Ophthalmoplegia R vision loss	Recovered
Cheng [3] 2012	72/M	R sphenoid sinus	R ptosis R vision loss	Partly improvement of visual acuity

Note: M: male; F: female; L: left; R: right; NLP: no light perception.

notably prostaglandins and collagenases [4]. The clinical symptoms of sinus mucocoeles are dependent on the region of the mass effect produced. There are many reports of ethmoid sinus mucocoeles causing optic neuropathy [4-10], however, ethmoid sinus mucocele causing orbital apex syndrome had rarely been reported. Orbital apex syndrome is typically used when all the structures at the orbital apex region and superior orbital fissure are affected by the disease process, which includes cranial nerves II, III, IV, V1 and VI [2]. The patient may manifest ptosis, proptosis, diplopia, ophthalmodynia, ophthalmoplegia, eye lid edema and visual loss.

There are only four cases of paranasal sinus mucocoeles including the present case that causing orbital apex syndrome in the English-language literature (**Table 1**). Efrat et al reported a case of onodi cell mucocele causing orbital apex syndrome, with prompt recovery after endoscopic removal. However, optic neuropathy did not improve and the patient remained blind [11]. Masaki et al reported a case of complete orbital apex syndrome due to sphenothmoid mucocele [2]. The patient underwent an endoscopic sinus surgery with cyst marsupialization. The eye symptoms gradually recovered, and visual acuity completely recovered 3 months after surgery [2]. Clarissa et al reported a

Orbital apex syndrome due to ethmoid mucocele

case of orbital apex syndrome secondary to a sphenoidal sinus mucocele and successfully treated with endoscopic drainage. One week after operation, there was a marked clinical improvement of visual acuity, ptosis and colour vision [3].

The diagnosis of ethmoid sinus mucocele may not be straightforward initially, especially if clinical manifestation is subtle or no proptosis. Patients usually do not have nasal symptoms, which make the diagnosis much more difficult. They may present to the neurologist with non-specific symptoms such as headache, as in our case, the major clinical manifestation was just headache initially, with no other typical symptoms.

Radiographic findings by CT and MRI are both important to identify ethmoid sinus mucocele and guide surgical management. Characteristic CT findings show an expansile, homogeneous mass with no contrast enhancement in the sinus [3]. CT scans provide insight into positioning and potential bony erosions caused by the expanding mucocele. The bone changes are best demonstrated by CT scans and can easily be overlooked on MRI scans. However, MRI is very useful to differentiate mucocèles from other lesions. On MRI, the appearance of mucocèles varies because of alterations in the protein concentration of the obstructed mucoid secretions. Depending on their biochemical constituents, mucocèles can be hypo-, iso-, or hyperintense or signal void on both T1- and T2-weighted images [1]. High signal intensity on T2-weighted and low intensity on T1-weighted MRI typically differentiate mucocèles from other masses, such as tumors. A higher protein content of mucocele mucus may increase the T1 signal intensity on MRI with contrast enhancement localized to the peripheral cystic walls [12].

The definitive treatment of choice for most mucocèles is based on prompt intranasal drainage and marsupialization using endoscopic sinus surgery. Early treatment reduced the threat of permanent visual loss and may result in recovery of vision. No clear duration of visual loss is predictive of visual outcome. Nonaka et al postulate that prognosis is related to the time interval between onset of symptoms and surgery, stating one month as a cutoff for poorer outcome [13].

In conclusion, ethmoid sinus mucocele causing orbital apex syndrome is extremely rare, the combination of CT and MRI imaging is the best choice for precise diagnosis and it should be immediately treated with early decompression and drainage of the cyst.

Acknowledgements

The present study was supported by the Health Department of Zhejiang Province, China (grant no. 2015116850), Science and Technology Department of Zhejiang Province, China (No. 2016C33144), and the National Natural Science Foundation of China (grant nos. 81172562 and 81372903).

Disclosure of conflict of interest

None.

Address correspondence to: Yang-Yang Bao, Department of Otolaryngology, The First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou 310003, Zhejiang Province, China. Tel: 86-571-87236894; Fax: 86-571-87236895; E-mail: bao4300383@yeah.net

References

- [1] Yong WW, Zhou SH, Bao YY. Sphenoid sinus mucocele presenting with oculomotor nerve palsy and affecting the functions of trigeminal nerve: a case report. *Int J Clin Exp Med* 2015; 15: 16854-168547.
- [2] Kumagai M, Hashimoto S, Suzuki H, Matsuura K, Takahashi E. Orbital apex syndrome caused by sphenothmoid mucocele. *Auris Nasus Larynx* 2003; 30: 295-297.
- [3] Cheng CS, Sanjay S, Yip CC, Yuen HW. Sphenoidal mucocele presenting as acute cranial nerve palsies. *Saudi J Ophthalmol* 2012; 26: 459-461.
- [4] Loo JL, Looi AL, Seah LL. Visual outcomes in patients with paranasal mucocèles. *Ophthal Plast Reconstr Surg* 2009; 25: 126-129.
- [5] Fukuda H, Fukumitsu R, Andoh M, Suzuki T, Yamana N, Kataoka H, Iwasaki K. Small Onodi cell mucocele causing chronic optic neuropathy: case report. *Neurol Med Chir (Tokyo)* 2010; 50: 953-955.
- [6] Wu W, Sun MT, Cannon PS, Jianbo S, Selva D. Recovery of visual function in a patient with an onodi cell mucocele compressive optic neuropathy who had a 5-week interval between onset and surgical intervention: a case report. *J Ophthalmol* 2010; 2010: 483056.

Orbital apex syndrome due to ethmoid mucocele

- [7] Toh ST, Lee JC. Onodi cell mucocele: rare cause of optic compressive neuropathy. *Arch Otolaryngol Head Neck Surg* 2007; 133: 1153-1156.
- [8] Yoon KC, Park YG, Kim HD, Lim SC. Optic neuropathy caused by a mucocele in an Onodi cell. *Jpn J Ophthalmol* 2006; 50: 296-298.
- [9] Yoshida K, Wataya T, Yamagata S. Mucocele in an Onodi cell responsible for acute optic neuropathy. *Br J Neurosurg* 2005; 19: 55-56.
- [10] Kitagawa K, Hayasaka S, Shimizu K, Nagaki Y. Optic neuropathy produced by a compressed mucocele in an Onodi cell. *Am J Ophthalmol* 2003; 135: 253-254.
- [11] Fleissig E, Spierer O, Koren I, Leibovitch I. Blinding orbital apex syndrome due to onodi cell mucocele. *Case Rep Ophthalmol Med* 2014; 2014: 453789.
- [12] Victores A, Foroozan R, Takashima M. Recurrent onodi cell mucocele: rare cause of 2 different ophthalmic complications. *Otolaryngol Head Neck Surg* 2012; 146: 338-339.
- [13] Nonaka M, Fukumoto A, Nonaka R, Ozu C, Baba S. A case of a mucocele in an Onodi cell. *J Nippon Med Sch* 2007; 74: 325-328.