Clinical Case Report

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Onodi cell mucocele causing isolated trochlear nerve palsy

A case report

Kyung Won Kwon, MD[a], Jin Seok Oh, MD[a], Ji Won Kim, MD[a], Roh-Eul Yoo, MD[b], Jae-Goo Kang, MD, PhD[a,*]

Abstract

Rationale: Acquired vertical diplopia is commonly observed in trochlear nerve palsy, often resulting from blunt head trauma or vascular problems. It is rarely caused by tumorous conditions or space occupying lesion. We report the first case of Onodi cell mucocele causing isolated trochlear nerve palsy.

Patient concerns: A 62-year-old male noticed a double vision which worsened when looking down. On ophthalmologic examinations, the patient showed no abnormalities in visual acuity, intraocular pressure, and no swelling in optic disc. In ocular motility test, he was notable for 10° left hypertropia in primary position, 6° in right head tilt position, 14° in left head tilt position and this was aggravated in right and down gaze. Ostiomeatal complex CT depicted an expansile soft tissue density completely filling the left Onodi cell.

Diagnoses: He was diagnosed with a trochlear nerve palsy caused by an isolated mucocele in the left Onodi cell.

Interventions: Three days after presentation, he underwent endoscopic sinus surgery for marsupialization of the mucocele in the left Onodi cell.

Outcomes: The mucocele was completely removed through the endoscopic endonasal approach. Within 4 months after surgery, his 4th nerve palsy had gradually and completely improved.

Lessons: Onodi cell mucoceles that cause trochlear nerve palsy are extremely rare. Timely surgical decompression is essential to achieve optimal recovery of the neural function. Combined trochlear nerve palsy should be evaluated when Onodi cell mucocele involves the orbital apex from above.

Abbreviation: OMC CT = ostiomeatal complex computed tomography.

Keywords: diplopia, endoscopic sinus surgery, Onodi cell mucocele, trochlear nerve palsy

1. Introduction

Trochlear nerve is the 4th cranial nerve that innervates the superior oblique muscle. Weakness of this muscle causes vertical double vision because the affected eye fails to perform downward gaze, often leading to its skewing in a slightly elevated position by an unopposed action of its antagonist muscle, the inferior oblique. The patient perceives two separate images projected onto different areas of his right and left retinae.

Approximately 50% of trochlear nerve palsy is presumed congenital, followed by hypertension (18%), trauma (18%). And it is rarely caused by tumorous conditions or space occupying lesion.[1] Mucoceles in the Onodi cell have not been reported as a cause of isolated trochlear nerve palsy.

Recently, the authors have experienced a case with Onodi cell mucocele causing vertical diplopia due to trochlear nerve palsy. The mucocele was completely removed through the endoscopic endonasal approach and the patient was gradually recovered from the paralysis. Here, we report its clinical features and progress with the review of the literatures. Informed written consent was obtained from the patient for publication of this case report and accompanying images.

2. Case report

A 62-year-old male with arrhythmia noticed a double vision which worsened when looking down starting 2 days before his visit to the emergency room in our hospital. Patient was healthy with no significant past medical or surgical history.

On ophthalmologic examinations, the patient showed no abnormalities in visual acuity and intraocular pressure. There was no swelling in optic disc, which might result from the optic neuritis. In ocular motility test, he was notable for 10° left

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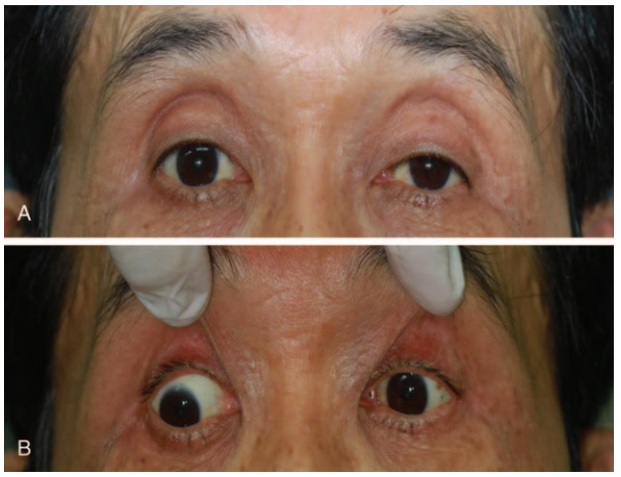

Figure 1. Clinical photograph of 62-year-old man with sudden onset diplopia showed 10° left hypertropia in primary position (A), which increased in right and down gaze (B).

hypertropia in primary position, 6° in right head tilt position, 14° in left head tilt position and this was aggravated in right and down gaze (Fig. 1). Patient did not experience pain on eye movement. Standard hematological investigations including C-reactive protein were normal.

The left trochlear nerve palsy was suspected and ostiomeatal complex computed tomography (OMC CT) was checked. OMC CT depicted an expansile soft tissue density completely filling the left Onodi cell (Fig. 2A and B, arrows). The lesion accompanied bony remodeling at the inferior wall of the left Onodi cell (Fig. 2B, arrowhead). The patient was transferred to our department and orbital magnetic resonance imaging (MRI) was performed for further characterization of the lesion.

On the orbital MRI, the lesion showed predominantly T1 hypointensity and T2 hyperintensity with focal T1 hyperintense and T2 hypointense foci (Fig. 3A and B, arrows). The inferior part of the lesion bulged into the lateral aspect of the optic nerve (Fig. 3D and E, arrowhead) where the left superior ophthalmic vein (Fig. 3E, empty arrowhead) was located. No demonstrable contrast enhancement was observed on the postcontrast T1-weighted image (Fig. 3C). No significant left superior oblique muscle atrophy was observed. We concluded that trochlear nerve palsy caused by an isolated mucocele in the left Onodi cell.

Three days after presentation, the patient underwent endoscopic sinus surgery for marsupialization of the mucocele in the left Onodi cell. Yellowish pus like discharge was observed intraoperatively and a thickened sac was removed. Histologically, chronic inflammation was found.

After surgery, the patient was treated with intravenous dexamethasone 10 mg/day, which was tapered to discontinuation after 1 week. After 2 weeks, the patient discharged with no postoperative complications, but with no improvement of diplopia. Within 4 months after surgery, his 4th nerve palsy had gradually and completely improved (Fig. 4). On ophthalmologic examinations, orthotropia primary position, orthotropia in right head tilt position, and 2° left hypertropia in left head tilt position. Postoperative ostiomeatal complex CT demonstrated that the left superior orbital fissure was successfully decompressed by total removal of the lesion (Fig. 5).

3. Discussion

Mucoceles are benign, encapsulated, expansile, locally destructive masses within sinus cavities, filled with mucous and lined by epithelium. Paranasal sinus mucoceles frequently involve the frontal sinus (65%), followed by the ethmoid (25%) and the maxillary sinus (10%). Only 1% to 2% of all paranasal sinus mucoceles are located in the sphenoid sinus or Onodi cell.[2,3]

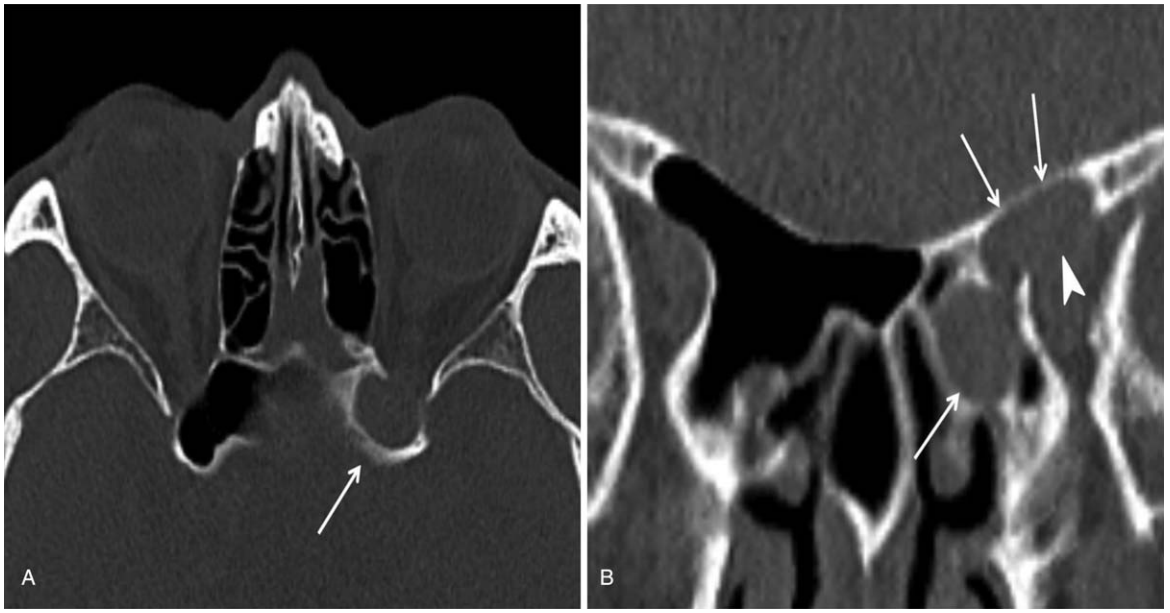

Figure 2. Ostiomeatal complex CT of the same patient. Axial (A) and coronal (B) scans showed an expansile soft tissue density completely filling the left Onodi cell (A and B, arrows). The lesion accompanied bony remodeling at the inferior wall of the left Onodi cell (B, arrowhead).

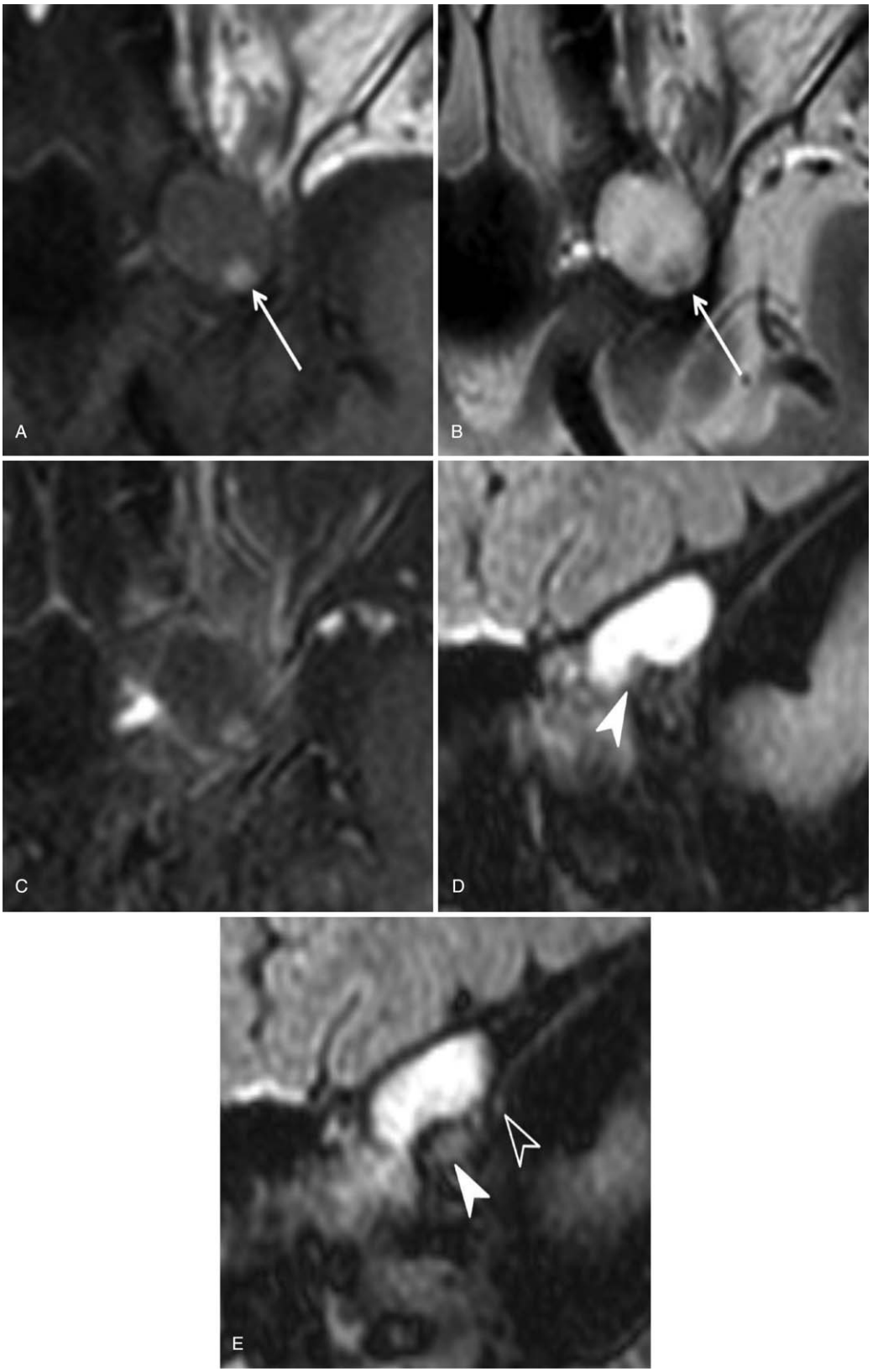

Figure 3. Orbital contrast MRI of the same patient. Axial scans showed predominantly T1 hypointensity (A) and T2 hyperintensity (B) with focal T1 hyperintense and T2 hypointense foci (A and B, arrows). No demonstrable contrast enhancement was observed on the postcontrast T1-weighted image (C). The inferior part of the lesion bulged into the lateral aspect of the optic nerve (D and E, arrowhead) where the left superior ophthalmic vein (E, empty arrowhead) was located.

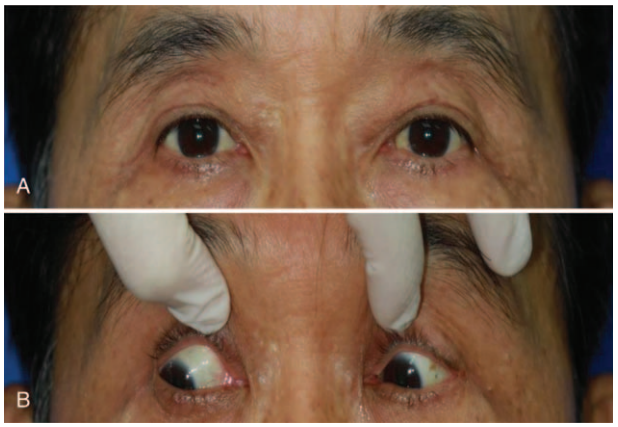

Figure 4. Clinical photographs of the same patient showed improvement of the left trochlear nerve palsy in 4 months later. Orthotropia in primary position (A), and right and down gazes (B).

An Onodi cell, is now defined as the most posterior ethmoid cell, which has pneumatized laterally and superiorly, and is intimately related to the optic nerve. The degree of pneumatization is variable and may lead to the Onodi cell encircling the optic nerve. Therefore, optic neuropathy is commonly occurred in the Onodi cell mucoceles.[4,5]

In published data, decreased visual acuity by compression of the optic nerve is the major and the only neurologic symptom of the Onodi cell mucocele. Contrarily, the 4th cranial nerve palsy was the presenting and solitary neurologic symptom in the current patient. After reviewing anatomical studies of the orbit and the trochlear nerve, the authors founded that there were clear anatomical reasons why the 4th cranial nerve was more vulnerable than the optic nerve in our case. The Onodi cell mucocele of our patient was centered above the orbital apex, exerting downward compression against the posterior orbital contents. Structures in the extraconal space (crainal nerve IV, frontal branch of the ophthalmic nerve, V1) were unprotected and vulnerable to the compression while intraconal structures (cranial II, III, VI) were guarded by the annulus of Zinn.

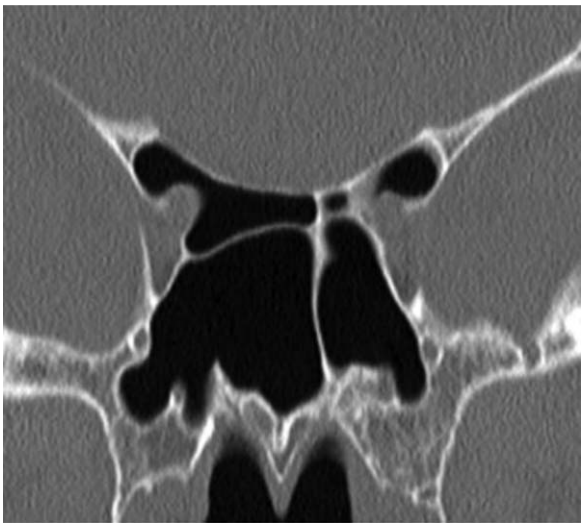

Figure 5. Postoperative ostiomeatal complex CT of the same patient. The left superior orbital fissure was successfully decompressed by total removal of the lesion.

According to current anatomical knowledge, upon entering the orbit from its cavernous course, the trochlear nerve makes a rather abrupt turn superomedially above the tendinous ring. The nerve continues its medial course, close to the roof of the orbit (above the levator palpebrae superioris muscle) to reach its target, the superior oblique muscle. Here, the nerve under the roof and over the tendinous ring is directly affected by the compressive force from above, hence it is especially bound to be vulnerable than the optic nerve and other intraconal structures. The optic nerve is the only cranial nerve which carries dural and arachnoid sheath along its course up to the globe. This provides additional protection from compression, which is deficient in case of the trochlear nerve. Therefore, it is necessary to scrutinize the possible trochlear nerve palsy when the mucocele involve the orbital apex from above: the trochlear palsy can be easily overshadowed by catastrophe of accompanying blindness.

Ischemic injury can be other possible cause of the trochlear nerve palsy of this patient. The vascular supply of the cranial nerve IV is entirely distinct from its neighboring cranial nerves such as III, V, and VI cranial nerves. While blood supply is mainly provided by the branches of internal carotid artery or the ophthalmic artery in the latter, vascular supply of the trochlear nerve is also contributed by the branches of the posterior ethmoid artery. If these posterior ethmoidal branches were the main contributor of arterial supply, ischemic nerve block could be a possible mechanism.[6,7,8]Inflammation may contribute to the trochlear nerve palsy. However, no pain on eye movement and abrupt onset of the palsy did not indicate that it played a causative role in this patient. Treatment of mucocele is based on prompt endoscopic surgical decompression and systemic steroids to reduce inflammation. Early intervention is very critical in recovery of neural function.[9,10] Controversy is still existed regarding postoperative recovery time after ocular motor nerve decompression. Improvement was usually seen within a week or two, occasionally up to a month according to the timing of surgical intervention.[10–12] Unlike previous reports, the course in our patient was much delayed by more than 4 months until complete recovery. Considering early surgical intervention (5 days after the onset of vertical diplopia), further studies are needed to determine the time course of neural recovery after surgical decompression.

Author contributions

Conceptualization: Kyung Won Kwon.
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Resources: Roh-Eul Yoo.
Supervision: Jae-Goo Kang.
Writing – original draft: Kyung Won Kwon.
Writing – review & editing: Ji Won Kim, Roh-Eul Yoo, Jae-Goo Kang.

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