Oculomotor Nerve Schwannoma: A Case Report

Yong-Hwan Cho1,2, Kyung-Su Sung1,2, Young-Jin Song1,2, Dae-Cheol Kim1,3, Sunseob Choi1,4, Ki-Uk Kim1,2

1Brain Tumor Institute Medical Science Research Center, Departments of 2Neurosurgery, 3Pathology, 4Radiology, College of Medicine, Dong-A University, Busan, Korea

Schwannomas account for about 8% of intracranial tumors and 90% are vestibular schwannomas. Oculomotor schwannoma without neurofibromatosis is extremely rare. A 41-year-old female presented with complaints of blurred vision, and the neurologic examination revealed afferent pupillary defect and decreased visual acuity of the left side. Brain magnetic resonance image showed an extra axial mass in the left superior orbital fissure. The patient underwent major surgery via the fronto-temporal approach. The tumor originated from the oculomotor nerve and was subtotally removed under microscopic surgery. The pathological findings confirmed the tumor as a schwannoma. After surgery, ptosis and medial gaze limitation of the left eye was detected, but the symptoms improved gradually.

Key Words Schwannoma; Oculomotor nerve; Surgery.

INTRODUCTION

Schwannomas are slowly growing peripheral nerve tumors that account for 6% to 8% of all intracranial tumors. They usually arise from the schwann cell layer of the vestibular branch of the eighth nerve or less commonly from the fifth nerve, the seventh nerve, and lower cranial nerve [1]. Oculomotor schwannoma without neurofibromatosis is very rare [2]. The first report of an isolated oculomotor nerve schwannoma was described by Kovacs in an autopsy in 1927 [3]. After then, approximately 40 cases of oculomotor nerve schwannomas have been described in the literature. The author reports a case of oculomotor schwannoma mimicking an optic nerve origin tumor that was removed surgically.

CASE REPORT

A 41-year-old female presented with one week history of blurred vision. A physical examination revealed relative afferent pupillary defect and decreased visual acuity of the left side. Visual acuity of the left eye was 20/200 with the Snellen’s chart (usual visual acuity was 200/200), and inferior hemi visual field defect of left eye was observed. The magnetic resonance imaging (MRI) revealed an avoid mass (24×8 mm), in the left superior orbital fissure. The lesion appeared as an iso-signal in the T1-weighted image, mild high signal in the T2-weighted image, and homogeneous enhancement in the gadolinium enhanced scan (Fig. 1A-C). The MRI showed that the small nodular mass, passing through the superior orbital fissure, extended from the cavernous sinus to the intraorbital region, and the mass was compressing the optic nerve. After administration of steroids, visual acuity improved, but did not fully recover. Thus, we decided to remove the tumor surgically, as this was an optic nerve originating mass with differential diagnostic possibilities. There was no evidence of neurofibromatosis, such as characteristic skin lesions, familial history, or other cranial nerve tumors or meningiomas in the MRI.

The patient underwent a major surgery via the left fronto-temporal approach using a neuro-navigation system. We performed an intradural anterior clinoidectomy and unroofing of the superior orbital wall. The tumor was observed to originate from the ventral portion of the oculomotor nerve (Fig. 2), and was removed subtotally. The anatomical continuity of the oculomotor nerve was preserved. Histopathological examination of the tumor revealed a schwannoma (Fig. 3). There was no evidence of recurrence on the one-year follow-up MRI (Fig. 1D-F). After surgery, the left side ptosis and medial gaze limitation was evident (Fig. 4A), but
Oculomotor Schwannoma

Fig. 1. A T1-weighted magnetic resonance imaging (MRI) with gadolinium enhancement is showing an avoid mass in the superior orbital fissure, and the mass was extended from cavernous sinus to the intraorbital resion (A-C). There is no evidence of re-growth on one year-following MRI (D-F).

Fig. 2. Intaoperative photogram showing the tumor originated from ventral part of oculomotor nerve. V1: ophthalmic branch of trigeminal nerve, III: oculomotor nerve, II: optic nerve, arrows: the tumor.

Fig. 3. The tumor showed whorling or wavy pattern of elongated spindle cells with nuclear palisading, alternating cellular and loose areas (H&E, ×200).

the symptoms improved gradually (Fig. 4B).

DISCUSSION

It is reported that intraorbital schwannomas account for 1–6% of all intraorbital tumors [2]. The accurate diagnosis, using imaging modalities, of oculomotor schwannoma may be difficult because of the complex orbital anatomy and its low incidence. Two clues leading to diagnosis are tumor location along the course of the oculomotor nerve, and oculo-
motor nerve palsy on neurologic examination [2,4]. The most common symptom of oculomotor schwannomas is oculomotor nerve palsy, but oculomotor nerve palsy is not always the initial symptom [4]. In the present case, optic nerve dysfunction was observed on neurologic examination without oculomotor nerve palsy. Thus, the optic nerve masses such as gliomas, schwannomas, or meningiomas appear to be differential preoperative diagnostic possibilities. However, we noted that the tumor origin was the oculomotor nerve during surgery. In patients with oculomotor schwannomas located in the intraorbital area without oculomotor nerve palsy, the operative finding may be important to confirm the tumor origin.

We reviewed 25 patients who received surgery for oculomotor nerve schwannomas (Table 1). Preoperative oculomotor nerve palsy manifested in 19 cases out of 25, and optic nerve dysfunction was shown in only 2 patients, including the present case. The tumor extended to the orbit region in 4 cases, and 2 of 4 patients manifested optic nerve dysfunction. Regardless of the radicality of the resection, postoperative oculomotor nerve palsy occurred in patients with oculomotor schwannomas in the orbital region, excluding our case.

Because of the tumor’s benign property, total resection of the tumor results in complete oculomotor nerve palsy [5-7]. Worsening of oculomotor nerve function may occur after subtotal or partial resection of the tumor [2,4]. The oculomotor nerve contains somatic motor fibers to many of the orbital muscles, but also carries parasympathetic fibers to the papillary muscle. Thus, oculomotor nerve palsy may worsen the patient’s quality of life. It is difficult to decide on the best treatment strategy for oculomotor schwannomas. According to each case, several treatment strategies have been reported by researchers. Katoh et al. [5] recommend ‘wait-and-see’ policy for asymptomatic patients with oculomotor schwannoma. According to Kim et al. [8], Gamma Knife radio-surgery may be an effective and minimally invasive treatment modality without risk of cranial nerve palsy in treatment of patients with schwannomas originating from the oculomotor, trochlear, and abducence nerves. Radical resection inevitably results in worsened oculomotor function, almost invariably in complete palsy. Thus, Asaoka et al. [4] recommend subtotal resection except large tumors that cause intractable symptoms. On the other hand, there were some reports of total resection of oculomotor schwannoma without permanent nerve palsy [3,9]. It’s location influences the radicality of tumor resection. The chance of oculomotor nerve injury after surgical resection may increase as the resection proceeds more anteriorly toward the superior orbital fissure [10]. In our review, past postoperative oculomotor nerve palsy.

![Fig. 4. Photograph demonstrating post-surgical function of oculomotor nerve. A: There were medial and down gaze limitation and ptosis of left eye. B: The nerve function was improved after four months.](image)
**Table 1.** Survey of oculomotor schwannomas treated by surgery in the literatures

| Author            | Age/Sex | Cranial nerve sign | Initial symptoms                                | Location                | Diameter (mm) | Resection            | Postoperative third nerve palsy |
|-------------------|---------|--------------------|------------------------------------------------|-------------------------|---------------|----------------------|---------------------------------|
| Okamoto et al. (1985) | 52/F    | III                | Exothlamos, seizure                             | Orbito-cisternocavernous | ND            | Subtotal resection    | Yes                             |
| Katsumata et al. (1990) | 47/M    | III                | Diplopia, ptosis                               | Cisternal               | 15            | Total resection       | ND                              |
| Lunardi et al. (1990) | 60/F    | III                | Hemiparesis, ptosis                            | Cisternal               | ND            | Total resection       | Yes                             |
| Mehta et al. (1990)    | 19/F    | III                | Gait disturbance                               | Cisternocavernous       | 50            | Subtotal resection    | Yes                             |
| Takano et al. (1990)    | 65/M    | III                | Ptosis, diplopia                               | Cavernous               | 25            | Partial resection     | Yes                             |
| Kurokawa et al. (1992) | 55/M    | III, V, VI         | Double vision                                  | Cavernous               | ND            | Total resection       | Yes                             |
| Kadota et al. (1993)    | 41/M    | III                | Ptosis, diplopia                               | Cisternal               | 20            | Total resection       | ND                              |
| Schulteiss et al. (1993) | 65/M    | No                 | Incidental                                     | Cisternal               | 8             | Total resection       | No                              |
| Niazi and Boggan (1994) | 13/M    | III                | Hemiparesis, diplopia, headache, dysarthria    | Cisternocavernous       | ND            | Subtotal resection    | Yes                             |
| Kachhara et al. (1998) | 55/F    | III, IV, V         | Headache, diplopia                             | Cavernous               | ND            | Total resection       | Yes                             |
| 61/F                | II, III, IV | Defective vision | Orbito-cisternocavernous                       | ND                      |               | Total resection       | Yes                             |
| Asaoka et al. (1999)    | 64/F    | No                 | Headache                                       | Cisternal               | 15            | Subtotal resection    | No                              |
| Mariniello et al. (1999) | 8/F     | III                | Diplopia                                       | Cavernous               | 10            | Total resection       | Yes                             |
| 66/F                | No      | Asymptomatic       | Cisternocavernous                              | ND                      |               | Subtotal resection    | Yes                             |
| Lingawi (2000)        | 53/M    | No                 | Headache                                       | Cisternal               | 5             | Total resection       | No                              |
| Sarma et al. (2002)    | 36/F    | III                | Diplopia                                       | Cavernous               | ND            | Total resection       | Yes                             |
| Hatakeyama et al. (2003) | 33/M    | III, V             | Diplopia, ptosis                               | Cisternocavernous       | 40            | Total resection       | No                              |
| Netuka et al. (2003)   | 12/F    | V                  | Headache                                       | Cisternal               | 28            | Total resection       | No                              |
| Ohata et al. (2006)    | 63/F    | III                | Diplopia, ptosis, eye pain, chemosis           | Orbito-cavernous        | 30            | Partial resection     | Yes                             |
| Tanriover et al. (2007) | 34/F    | III                | Anisocoria, ptosis, headache, exotropia        | Cisternal               | 20            | Subtotal resection    | No                              |
| Prabhu and Bruner (2009) | 38/F    | III                | Headache, diplopia, ptosis, dizziness          | Cisternocavernous       | 35            | Total resection       | Yes                             |
| Goel and Shah (2010)   | 32/M    | III                | Headache, diplopia, ptosis                     | Cisternal               | ND            | Subtotal resection    | No                              |
| 16/M                | III, IV, V | Headache, ptosis | Cisternocavernous                              | ND                      |               | Total resection       | Yes                             |
nerve palsies occurred in all patients with tumors that extended to the orbital region, except our present case (Table 1), where the tumor was located in the superior orbital fissure and close to other neurovascular structures. Also, the preoperative oculomotor nerve function was preserved. Thus, we decided to perform a subtotal resection to avoid complete oculomotor nerve palsy, and planned an adjuvant frameless radiosurgery for the remnant tumor. During the operation, most of the mass was removed leaving the tumor capsule. In the postoperative MRI, the target lesion could not be detected so we decided not to perform the adjuvant radiosurgery. There is no evidence of tumor recurrence on the one-year follow-up MRI, and we planned for follow-up MRI annually.

In conclusion, we report a case of oculomotor schwannoma in which the preoperative diagnosis may have been very difficult due to the complex orbital anatomy and the low incidence of disease. The treatment strategy should be established considering the preoperative nerve functions and the tumor location. We removed the tumor subtotally using the fronto-temporal approach with resulting temporary oculomotor nerve palsy.

**Conflicts of Interest**

The authors have no financial conflicts of interest.

**REFERENCES**

1. Choi YS, Sung KS, Song YJ, Kim HD. Olfactory schwannoma-case report-. J Korean Neurosurg Soc 2009;45:103-6.
2. Ohata K, Takami T, Goto T, Ishibashi K. Schwannoma of the oculomotor nerve. Neurol India 2006;54:437-9.
3. Netuka D, Benes V. Oculomotor nerve schwannoma. Br J Neurosurg 2003;17:168-73.
4. Asaoka K, Sawamura Y, Murai H, Satoh M. Schwannoma of the oculomotor nerve: a case report with consideration of the surgical treatment. Neurosurgery 1999;45:630-3; discussion 633-4.
5. Katoh M, Kawamoto T, Ohnishi K, Sawamura Y, Abe H. Asymptomatic schwannoma of the oculomotor nerve: case report. J Clin Neurosci 2000;7:458-60.
6. Mehta VS, Singh RV, Misra NK, Choudhary C. Schwannoma of the oculomotor nerve. Br J Neurosurg 1990;4:69-72.
7. Lunardi P, Rocchi G, Rizzo A, Missori P. Neurinoma of the oculomotor nerve. Clin Neurol Neurosurg 1990;92:333-5.
8. Kim IY, Kondziolka D, Niranjan A, Flickinger JC, Lunsford LD. Gamma Knife surgery for schwannomas originating from cranial nerves III, IV, and VI. J Neurosurg 2008;109 Suppl:149-53.
9. Schultheiss R, Kristof R, Schramm J. Complete removal of an oculomotor nerve neurinoma without permanent functional deficit. Case report. Ger J Ophthalmol 1993;2:228-33.
10. Tanriover N, Kemerdere R, Kafadar AM, Muhammedrezai S, Akar Z. Oculomotor nerve schwannoma located in the oculomotor cistern. Surg Neurol 2007;67:83-8; discussion 88.