Case Report

Oculomotor Nerve Schwannoma: A Case Report and Review of the Literature

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Background: *Oculomotor nerve schwannomas are extremely rare tumors. There are only 40 cases reported in the literature. There is no standard treatment for these rare tumors.*
Case Report: *The authors have reported a case of a 41-year-old Thai man presenting with progressive visual loss of the left eye for 6 months without diplopia. Visual acuity was 20/70 in the right and 20/400 in the left. There was no limitation of eye movement. MRI showed a 42.5 ml mass in the suprasellar region compatible with a schwannoma. The patient underwent a left pterional craniotomy with partial tumor removal. The pathological section confirmed a diagnosis of schwannoma and the patient received postoperative stereotactic radiotherapy.*
Conclusion: *Options for treating these rare tumors include clinical observation, surgical resection or stereotactic radiation. High incidence of complete third nerve palsy following surgery has been reported in the literature. Therefore, a subtotal removal of large oculomotor schwannoma followed by stereotactic radiotherapy could provide a safer alternative compared to radical surgery.*

Keywords: *Oculomotor nerve, Schwannoma*

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Intracranial schwannomas account for 8% of brain tumors. Most of intracranial schwannomas originate from the sensory nerve. The most common cranial nerve that affected by schwannoma is the vestibulocochlear nerve (CN.VIII), followed by trigeminal nerve. Motor nerve schwannoma is very rare in the absence of neurofibromatosis. There are only about 40 cases of oculomotor schwannomas that have so far been reported in the literatures. The authors present an additional case of this rare tumor.

Case Report

A 41-year-old Thai man presented with progressive visual loss of his left eye for 6 months without diplopia. He also complained of dull headache and left orbital pain. General physical examination was normal. There were no neurocutaneous markers. Visual acuity in the right eye was 20/70 and in the left eye was 20/400. The right pupil, 3 mm, reacted to light. The left pupil, 4 mm, also reacted to light. Relative afferent pupillary defect (RAPD) presented in the left eye. There was a constriction of visual field in the left eye. Fundoscopic examination revealed a pale optic disc in the left eye. The visual field and fundus of the right eye were normal. There was no ptosis and no limitation of eye movement in any direction in either eye. Neurological examination revealed no facial numbness, no facial weakness and no motor weakness. Other neurological examinations were within normal limits.

Routine investigations were normal. Magnetic resonance imaging (MRI) showed a 42.5 ml large inhomogeneous iso/hyposignal T1 and inhomogeneous intermediate and marked hypersignal T2 with mixed solid and cystic enhancing suprasellar mass extending into the left cavernous sinus severely compressing the anterior pons and midbrain posteriorly, third ventricular floor superiorly and anterior and upwardly displacing optic chiasm, highly suggestive of left oculomotor nerve schwannoma and less likely meningioma and optic glioma (Fig 1A-C). From the physical examination and MRI, the cause of visual

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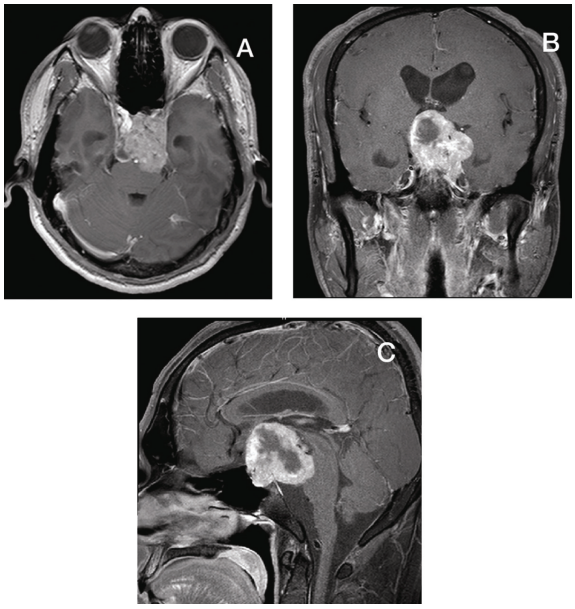

Fig. 1 Pre-operative MRI, A) axial, B) coronal and C) sagittal view

impairment in the left eye was most likely from left optic nerve compression by the tumor mass.

The patient was operated on on June 5, 2009. Left pterional approach craniotomy and subtotal excision of the tumor were achieved. At operation, left sphenoid wing was drilled, sylvian fissure was opened; then a large soft mixed solid and cystic tumor was found. Tumor was internally debulked and sent for frozen section. Pathological result revealed a schwannoma. Tumor was further removed in piecemeal fashion. Left oculomotor nerve was not able to be identified at the beginning of the operation. It was markedly attached to the posterior part of the tumor and it was thought that the tumor originated from it. Unfortunately, the left oculomotor nerve and left anterior cerebral artery (ACA) were injured at internal carotid artery (ICA) bifurcation during tumor resection. It was believed to result from prolonged brain retraction. The authors had tried to suture at the laceration site, but it failed because the defect was mainly located at the posterior wall of internal carotid bifurcation. The authors decided to apply a clip at internal carotid artery bifurcation after confirming that the right A1 segment of the right anterior cerebral artery was large enough.

The patient had complete left oculomotor nerve palsy and right hemiparesis with motor power grade 4 immediately after the operation. At 2 days postoperation, the patient developed progressive weakness on the right side with motor power about grade 2 without dysphasia. Computerized tomography (CT) scan showed infarction of left basal ganglia (Fig. 2).

Permanent section pathological result showed characteristics of schwannoma (Fig. 3). Post-operative MRI 4 months after surgery revealed residual tumor at left cavernous sinus and left sided prepontine cistern, measuring 5.4 ml. Optic pathway compression is not seen (Fig 4A-C). The patient underwent stereotactic radiotherapy between November 25 and December 1, 2009 by cyberknife. At 7-month follow-up his right

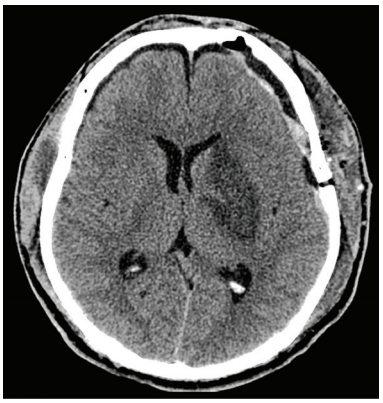

Fig. 2 Post-operative CT scan 2 days after surgery showed infarction of left basal ganglia

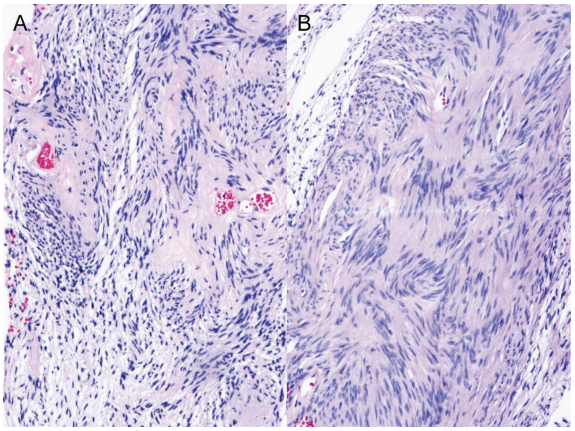

Fig. 3 Pathological section. A) The low magnification of Schwannoma shows swirls of compact Antoni A and loose Antoni B tissue with a generally prominent vascular pattern (H&E, x100), B) The section shows Verrocay bodies consisting of palisades of cells separated by pink-stained zones composed of cell processes (H&E, x100)

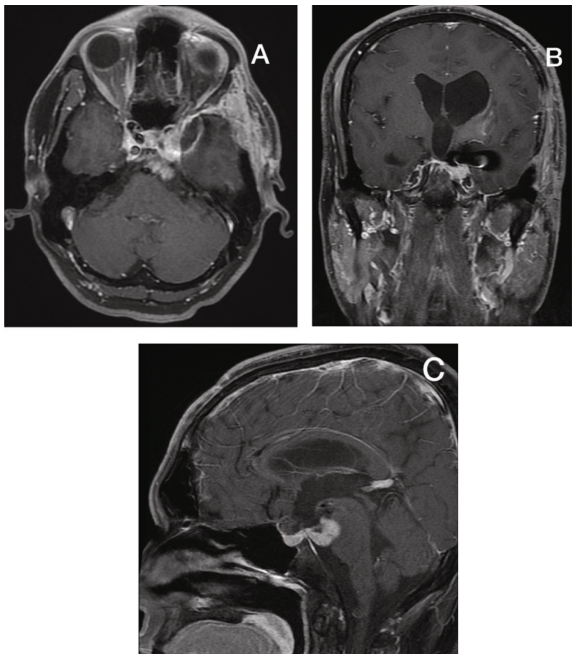

Fig. 4 Post-operative MRI A) axial, B) coronal and C) sagittal view

hemiparesis had improved with motor power about grade 4. He could walk and took care of himself without support. He still has complete right oculomotor nerve palsy. The visual deficit has improved. Visual acuity in the right eye is 20/30 and the left eye is 20/200.

Discussion

Schwannomas usually occur in the sensory nerve. Schwannomas of motor nerve are very rare without neurofibromatosis. The first case report of oculomotor schwannoma was by Kovacs[1]. He reported a case from autopsy in 1927. Since then, there have been about 40 cases of oculomotor schwannoma reported in the literature. The first case from Thailand was reported by Shuangshoti[2] in 1975. He reported a case of left oculomotor schwannoma that was accidentally found during autopsy. Review of the literature on oculomotor nerve schwannomas are summarized in Table 1.

Tumor diameter was 45 mm in our case. Tumor size varies from 4 to 55 mm in the literature. Most of the cases have oculomotor nerve deficit before surgery. Extent of resection varies from partial to total tumor removal. In earlier cases, tumors were relatively large and postoperative oculomotor nerve palsy was almost always the result[3-5,7,9,11].

Three cases in the literature have right hemiparesis. The tumors were described as large tumors in 2 cases[3,6]. Shuangshoti[2] reported a case of tumor 5 mm in diameter with right hemiparesis. He described that there was a fusiform aneurysm at left ICA with severe atherosclerosis, which might be the cause of right hemiparesis.

There were 2 cases in the literature that underwent total tumor removal with nerve grafting[8,10]. Oculomotor nerve functions were partially improved in both cases. These may be from several functions of oculomotor nerve including eye opening (levator palpebrae muscle), eye movement (medial rectus, superior rectus, inferior rectus and inferior oblique muscle) and tupillary constriction that limit the outcomes of nerve grafting.

There were 2 case reports of malignant nerve sheath tumor of oculomotor nerve[13,19]. Both cases are not associated with neurofibromatosis. Kozic et al[13] reported a 9-year-old boy with malignant peripheral nerve sheath tumor of oculomotor nerve. MR spectroscopy revealed the presence of a high choline peak with no detectable concentrations of creatine and N-acetyl aspartate in the mass. The patient underwent partial tumor removal but the clinical condition deteriorated from aggressive progression of the tumor growth. Sener suggested that diffusion MRI could aid in differentiating malignant from benign oculomotor nerve schwannoma[19]. Malignant oculomotor nerve schwannoma is hyperintense on b = 1000 sec/mm^2 images, and ADC values are low compared to the isointensity, and high ADC values of benign schwannoma.

Bisdorff et al[14] reported a 14-year-old girl with oculomotor nerve schwannoma mimicking ophthalmoplegic migraine. She had right periorbital headache of pressing and pulsating character. By the end of headache she developed a complete ipsilateral oculomotor nerve dysfunction lasting for 1-2 months with a slow complete recovery. The frequency of the episodes was between 2-3 per year and every 1-2 years. MR image revealed nodular lesion of 7 mm diameter on the cisternal portion of right oculomotor nerve with the same size as a previous MRI performed 10 years earlier.

Chewning et al[17] reported false positive diagnosis of an intracranial aneurysm using 3D CT angiography. The patient was a 3-year-old girl with right oculomotor nerve palsy with papillary involvement. 3D CT angiography revealed an aneurysm at right P1-P2 junction of the right posterior

Table 1. Summary of reported cases of oculomotor schwannoma

	Age/ sex	Oculomotor nerve signs	Other signs	Neurofibromatosis	Diameter (mm)	Extent of resection	Postoperative course
Kovacs[1]	55/M	None	None	ND	22	(autopsy)	ND
Shuangshoti[2]	64/F	Ptosis, dilated pupil, limitation of inward eye rotation	Right hemiparesis, headache	ND	5	(autopsy)	ND
Leunda et al[3]	11/M	Impairment of upward gaze in left eye	Right hemiparesis, headache	ND	55	Total	CN III palsy
Takano et al[4]	65/F	Ptosis, dilate pupil, impairment of downward, upward and inward gaze	None	Absence	25	Partial	CN III palsy
Kurokawa et al[5]	55/M	Oculomotor paralysis	CN VI paresis, hypesthesia on right face	Absence	ND	Total	CN III palsy
Niazi et al[6]	13/M	Ptosis, slight anisocoria and impairment of inward gaze	Right facial weakness Right hemiparesis	ND	Large	Total	Improved CN III function and hemiparesis
Kachhara et al[7]	55/F	Ptosis, diplopia	CN IV palsy, hypesthesia in ophthalmic division of CN V	Absence	ND	Total	CN III IV and VI palsy, hypesthesia in ophthalmic division of CN V
	61/M	Ptosis, dilated pupil, impairment of inward gaze	Proptosis, CN IV palsy	Absence	ND	Total	CN III and IV palsy
Mariniello et al[8]	8/F	Ptosis, dilated pupil, impairment of downward, upward and inward gaze	Proptosis	Absence	ND	Total with nerve grafting	Improved ptosis, downward and upward eye movement
Katoh et al[9]	66/F	None	None	ND	ND	Partial	CN III palsy
Sarma et al[10]	ND	Diplopia	None	ND	ND	Total with nerve grafting	Improved CN III function
Netuka et al[11]	12/F	None	None	Absence	28	Total	CN III palsy
Hatakeyama et al[12]	33/M	Ptosis, dilated pupil, impairment of downward, upward and inward gaze	Hypesthesia in maxillary and mandibular division of CN V	Absence	40	Total	Improved CN III and V function
Kozic et al[13]	9/M	Ptosis	None	Absence	27	Partial	Deteriorated clinical condition
Bisdorff et al[14]	14/F	Ptosis, dilated pupil, impairment of downward, upward and inward gaze	None	ND	7	None	Improved CN III function
Tanriover et al[15]	34/F	Ptosis, dilated pupil, impairment of inward gaze	Pale optic disc	ND	ND	Partial	Improved CN III function
Kim et al[16]	29/M	ND	None	ND	ND	Partial	Ptosis
	19/F	Ophthalmoplegia	None	ND	ND	None	Same
Chewning et al[17]	3/F	Ptosis, dilated pupil	None	ND	4	None	Same
Nishioka et al[18]	ND	ND	ND	Absence	10	Partial	ND
Sener[19]	1/M	Ptosis, dilated pupil, impairment of downward, upward and inward gaze	None	Absence	30	Total	ND
Saetia et al	41/M	None	Progressive visual loss	Absence	45	Partial	CN III palsy, right hemiparesis

CN II: optic nerve, CN III: oculomotor nerve, CN IV: trochlear nerve, CN V: trigeminal nerve, CN VI: abducens nerve, ND: not described

cerebral artery. Subsequent digital subtraction angiography did not show any aneurysm. MRI revealed a 4 mm oculomotor nerve schwannoma.

Treatment of large oculomotor nerve schwannoma with mass effect to surrounding structure is surgical resection but that of small tumors is still controversial. Most schwannomas of CN.V and CN.VIII can be controlled with stereotactic radiosurgery or stereotactic radiotherapy. There are anecdotal reports about the role of these treatment modalities for oculomotor nerve schwannoma. Kim et al[16] reported 2 cases of oculomotor schwannoma who received gamma knife radiosurgery. The cases were followed for 9 and 36 months respectively without clinical worsening and tumor progression. Nishioka et al[18] reported 1 case of oculomotor nerve schwannoma who received stereotactic radiotherapy. The tumor size was decreased and the patient had no clinical worsening. From these reports, stereotactic radiation may have roles in small oculomotor schwannomas or residual tumors after surgical resection. Unfortunately, the number of cases is small. Further investigations are needed to conclude about standard treatment for oculomotor nerve schwannoma.

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รายงานผู้ป่วยเนื้องอกชวานโนม่าของเส้นประสาทสมองคู่ที่สาม

เกรียงศักดิ์ แซ่เตีย, นพดล ลาภเจริญทรัพย์, ณรงเดช เวชกามา

***ภูมิหลัง**: เนื้องอกชวานโนม่าของเส้นประสาทสมองคู่ที่สามพบได้น้อยมาก มีรายงานทั่วโลกเพียง 40 ราย เท่านั้น โดยไม่มีแนวทางการรักษาที่เป็นมาตรฐานสำหรับเนื้องอกที่พบน้อยชนิดนี้*

***รายงานผู้ป่วย**: ผู้ป่วยชายไทย อายุ 41 ปี มีอาการตาซ้ายมัวลงภายใน 6 เดือน ไม่มีอาการเห็นภาพซ้อนระดับสายตาข้างขวา 20/70 ข้างซ้าย 20/400 การกลอกตาเป็นปกติ ผลการตรวจสมองด้วยคลื่นแม่เหล็กไฟฟ้า พบก้อนเนื้องอกขนาด 42.5 มิลลิลิตร ที่บริเวณเหนือต่อเซลล่า ซึ่งเข้าได้กับเนื้องอกชนิดชวานโนม่า ผู้ป่วยได้รับการผ่าตัดเนื้องอกสมองดังกล่าว และผลทางพยาธิวิทยายืนยันการวินิจฉัยว่าเป็นเนื้องอกชวานโนม่า ผู้ป่วยได้รับการรักษาต่อด้วยการฉายรังสี*

***สรุป**: ทางเลือกของการรักษาเนื้องอกชนิดนี้ มีทั้งการเฝ้าติดตามอาการ การผ่าตัดรวมไปถึงการฉายรังสี พบอุบัติการณ์สูงว่ามีเส้นประสาทสมองคู่ที่สามเป็นอัมพาตหลังการผ่าตัดที่เอาเนื้องอกออกทั้งหมด ดังนั้นในรายที่เนื้องอกขนาดใหญ่ การผ่าตัดเอาเนื้องอกออกบางส่วนแล้วฉายแสงต่อนับเป็นทางเลือกที่ปลอดภัยกว่า*