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Oculomotor Nerve Schwannoma Associated with Acute Hydrocephalus: Case Report

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Abstract

A 37-year-old woman presented with an extremely rare large oculomotor schwannoma associated with acute hydrocephalus manifesting as semicoma and anisocoria. Brain computed tomography and magnetic resonance imaging revealed a tumor in the oculomotor cistern. Cerebral angiography revealed separation of the posterior cerebral artery (PCA) and superior cerebellar artery (SCA). The tumor was removed subtotally by two stage surgery. Histological examination revealed ordinary schwannoma. The diagnosis of oculomotor nerve schwannoma was based on the intraoperative finding of the tumor origin in the oculomotor nerve. Oculomotor nerve schwannoma can cause acute hydrocephalus and manifest as impaired consciousness. The angiographical separation of the PCA and SCA was very useful for the preoperative diagnosis of oculomotor nerve schwannoma.

Key words: oculomotor nerve, schwannoma, angiography, magnetic resonance imaging

Introduction

Schwannomas arising from the oculomotor nerves are rare, unless associated with neurofibromatosis. Isolated oculomotor nerve schwannoma was first observed during an autopsy in 1927.[1,2] Large oculomotor schwannoma is extremely rare, because this tumor usually manifests as oculomotor palsy whilst still small. Only 28 cases of surgical resection of oculomotor nerve schwannoma have been reported.[2–27] Here we describe a case of large oculomotor nerve schwannoma associated with acute hydrocephalus manifesting as semicoma and anisocoria.

Case Report

A 37-year-old woman was transferred to our hospital with head injury due to falling. She had been in the mental hospital for 3 months because of abnormal behavior. Routine examinations in the emergency room found no abnormalities. Neurological examination detected cognitive impairment and anisocoria (right 4.5 mm, left 2.5 mm). The left pupil was promptly reactive to light, but the right pupil was not reactive. Examination of voluntary eye movement was not possible because she was drowsy and disoriented. Her consciousness level was 8 on the Glasgow Coma Scale (E2 V2 M4). Computed tomography revealed an acute epidural hematoma in the left convexity and a mass lesion in the right middle cranial fossa extending to the cerebellopontine angle. Magnetic resonance imaging showed a tumor (50 × 37 × 40 mm) in the middle cranial fossa extending to the cerebellopontine angle, containing multiple intratumoral cysts, with heterogeneous enhancement after gadolinium injection (Fig. 1). The third ventricle was displaced superiorly. Both lateral ventricles were enlarged, indicating acute hydrocephalus. Cerebral digital subtraction angiography revealed that the posterior cerebral artery (PCA) was displaced superiorly and the superior cerebellar artery (SCA) was displaced inferiorly. The lesion was avascular (Fig. 2).

The acute hydrocephalus was considered to be caused by an obstruction of the third ventricle and aqueduct. Semi-emergency tumor resection was performed through the right pterional route. The tumor was xanthochromic, elastic, and soft, and did not bleed easily. Half of the tumor was resected. The tumor originated from the oculomotor nerve, and the part of the tumor involving the oculomotor nerve was left. Consequently, the oculomotor nerve was preserved anatomically. The caudal part of the tumor was not visible and could not be resected through this approach (Fig. 3A). Postoperatively, she continued to

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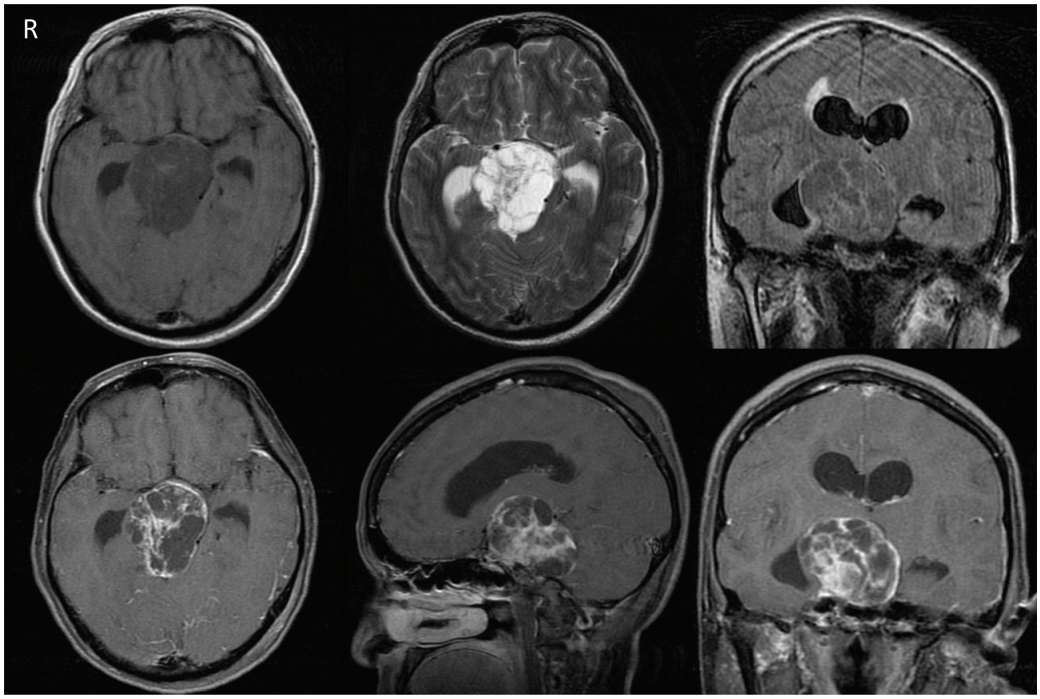

Fig. 1 T_1-weighted (*upper left*), T_2-weighted (*upper center*), fluid-attenuated inversion recovery (*upper right*), and T_1-weighted with gadolinium (*lower row*) magnetic resonance images at the initial presentation showing a large ring-enhanced cystic lesion in the right parasellar region, and a heterogeneously enhanced lesion extending to the posterior cranial fossa with displacement of brain stem structures and the region of the third ventricle.

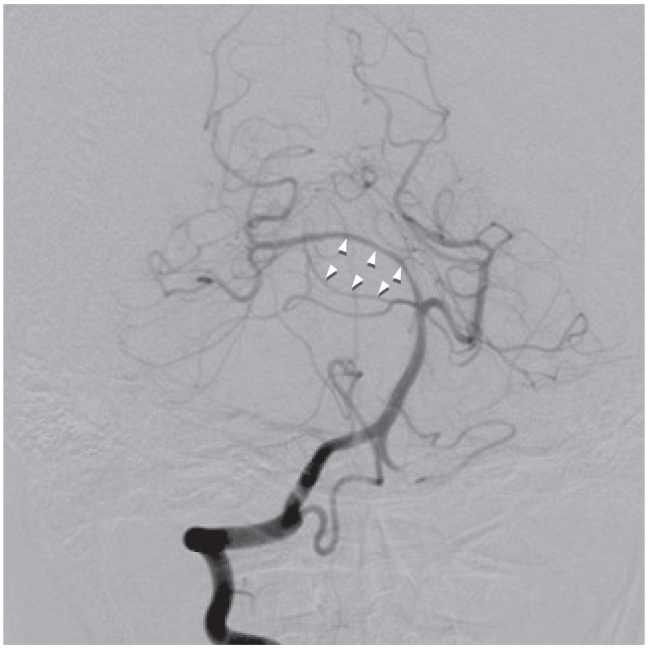

Fig. 2 Preoperative cerebral digital subtraction angiogram of the posterior circulation indicating separation of the posterior cerebral artery and superior cerebellar artery by the mass lesion (*white arrowheads*).

have complete third nerve paralysis on the right, but the acute hydrocephalus improved. Histological examination confirmed a diagnosis of schwannoma (Fig. 4). Six weeks later, second operation to remove the remnant tumor was performed through the right subtemporal route (Fig. 5). The tumor was subtotally removed (Fig. 3B). Her general condition gradually improved and neurological condition fully recovered including the complete right oculomotor nerve paralysis after rehabilitation for 2 months.

Discussion

Only 28 cases of surgical resection of oculomotor nerve schwannoma have been reported (Table 1).[2–27] Including our patient, there were 15 females and 14 males aged from 8 to 66 years (mean age 40.0 years). The most common initial symptom of oculomotor nerve schwannoma was oculomotor palsy (16 patients) followed by headache (12 patients). Semicoma has not been reported as an initial symptom of oculomotor nerve schwannoma. No emergent case of large oculomotor schwannoma associated with acute hydrocephalus has been reported. Among the 29 patients with oculomotor nerve schwannoma, 25 patients (86%) had oculomotor nerve palsy preoperatively. Among these 25 patients, 17 patients suffered oculomotor nerve palsy postoperatively, 4 patients had improved oculomotor

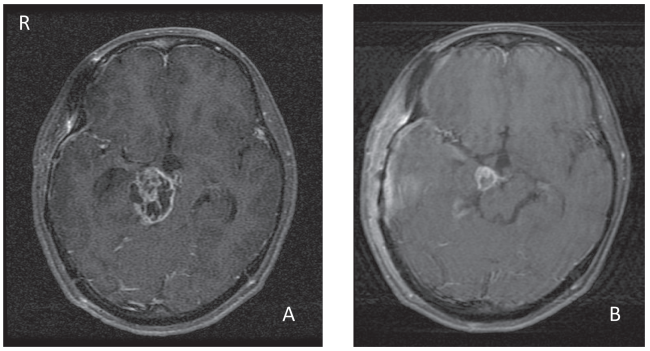

Fig. 3 T_1-weighted with gadolinium magnetic resonance images after the first (A) and second (B) operations showing tumor remains in the oculomotor cistern.

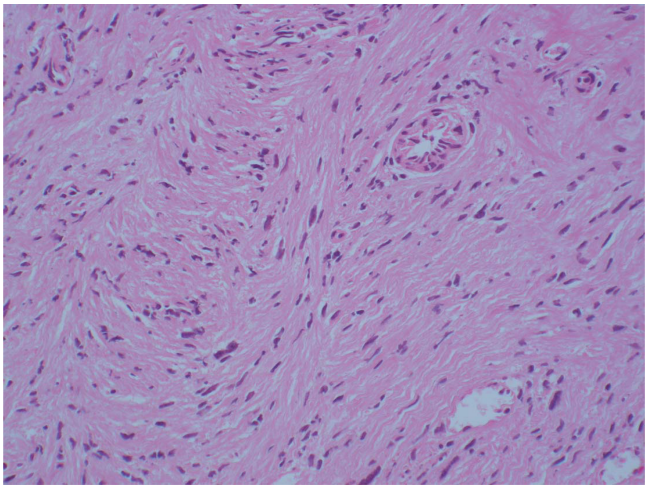

Fig. 4 Photomicrograph of the resected lesion demonstrating a cellular pattern consistent with schwannoma. Hematoxylin and eosin stain, original magnification ×40.

nerve palsy, and 4 patients had no described postoperative neurological state. Therefore, the oculomotor nerve was preserved in 4 of 25 patients (16%). Consequently, oculomotor nerve palsy is likely to occur in most patients after treatment despite anatomic preservation of the oculomotor nerve.[22] Only 2 patients had symptoms of increased intracranial pressure preoperatively. Even if small, oculomotor nerve schwannoma is very likely to manifest as oculomotor nerve palsy. Consequently, most patients have undergone imaging examination before the tumor can become large.

In our review, the origin of the schwannoma was identified by intraoperative finding in 18 cases. The diagnosis was based on the operative, neurological, and histological findings in the other 10 cases. Preoperative diagnosis of oculomotor nerve schwannoma is difficult mainly because of its low incidence and nonspecific radiological features.[26,27] Preoperative diagnosis was discussed in 14 previous cases, and oculomotor schwannoma was suspected in only two cases. In these two cases, the diagnostic reason was an enlargement of the superior orbital fissure on plain craniogram because the tumor extended into the orbital cavity,[21] and the presence of preoperative anisocoria.[23] Anisocoria is not a specific symptom in neurosurgery patients, so does not strongly indicate oculomotor nerve schwannoma, especially in the presence of hydrocephalus as in our case. In our case, preoperative cerebral digital subtraction angiography clearly demonstrated separation of the PCA and SCA. The oculomotor nerve normally passes between the PCA and SCA so that such separation of these vessels strongly supported the diagnosis of oculomotor nerve schwannoma.[19] Only one previous case showed separation of the PCA and SCA.[19] In that

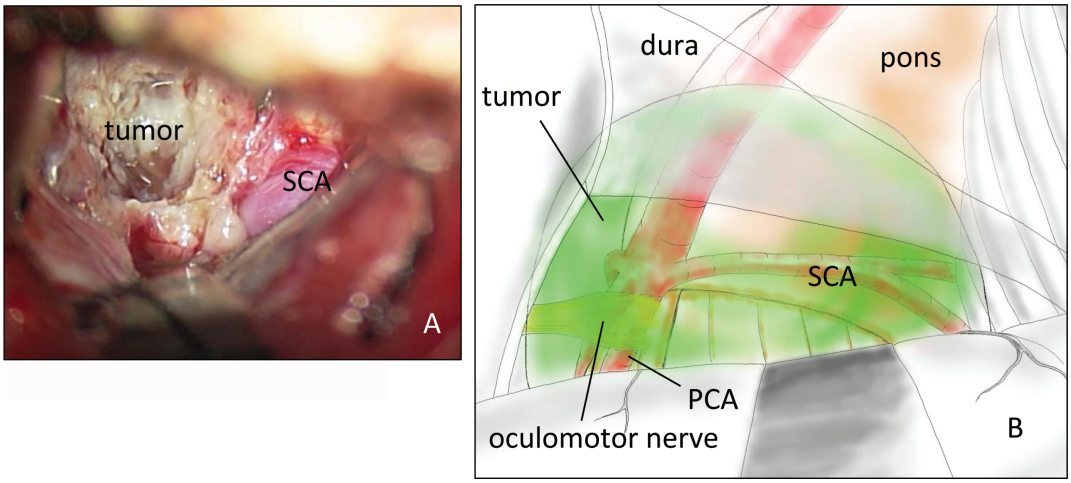

Fig. 5 A: Intraoperative photograph showing the tumor and the superior cerebellar artery (SCA), with the oculomotor cistern filled with tumor. B: Drawing showing the anatomical relationship between the tumor and the posterior cerebral artery (PCA), SCA, and oculomotor nerve. The oculomotor nerve is spread and involved in the tumor. The PCA is displaced superiorly and the SCA is displaced inferiorly.

Table 1 Cases of oculomotor nerve schwannoma treated by surgery

Case No.	Author	Year	Age (yrs)	Sex	Initial symptoms	Cranial nerve sign	Diameter (mm)	Resection	Third cranial nerve palsy	
									Preoperative	Postoperative
1	Schubiger et al.[25]	1980	19	F	Headache	III-V	30	Total	Yes	No (improved)
2	Broggi and Franzini[4]	1981	45	M	Hemiparesis	III	30	Total	Yes	ND
3	Hiscott and Symon[7]	1982	58	F	Hemiparesis, dementia	III	40	Subtotal	Yes	Yes
4	Leunda et al.[14]	1982	11	M	Headache, hemiparesis	III, IV	55	Total	Yes	Yes
5	Kansu et al.[10]	1982	15	M	Headache	III	4	Total	Yes	ND
6	Okamoto et al.[21]	1985	52	F	Diplopia	III	40	Subtotal	Yes	Yes
7	Katsumata et al.[12]	1990	47	M	Diplopia, ptosis	III	15	Total	Yes	ND
8	Lunardi et al.[15]	1990	60	F	Headache	III	35	Total	Yes	Yes
9	Mehta et al.[17]	1990	19	F	Gait disturbance	III	50	Subtotal	Yes	Yes
10	Takano et al.[27]	1990	65	M	Ptosis, diplopia	III	25	partial	Yes	Yes
11	Kurokawa et al.[13]	1992	55	M	Diplopia	III, V, VI	20	Total	Yes	Yes
12	Kadota et al.[9]	1993	41	M	Diplopia, ptosis	III	20	Total	Yes	ND
13	Schultheiss et al.[26]	1993	65	M	Incidental	None	8	Total	No	No
14	Niazi and Boggan[19]	1994	13	F	Hemiparesis, diplopia, headache, dysarthria	III	30	Total	Yes	Yes
15	Kachhara et al.[8]	1998	55	M	Headache	III, V	20	Total	Yes	Yes
16			61	M	Diminished vision	III, IV	40	Total	Yes	Yes
17	Asaoka et al.[3]	1999	64	F	Headache	None	15	Subtotal	No	No
18	Mariniello et al.[16]	1999	8	F	Diplopia	III	10	Total	Yes	Yes
19	Katoh et al.[11]	2000	66	F	None	None	15	Partial	No	Yes (deteriorated)
20	Sarma et al.[24]	2002	36	F	Diplopia	III	10	Total	Yes	Yes
21	Hatakeyama et al.[6]	2003	33	M	Diplopia	III, V	40	Total	Yes	No (improved)
22	Netuka and Benes[18]	2003	12	F	Headache	II,V	28	Total	No	No
23	Ohata et al.[20]	2006	63	F	Diplopia, ptosis, chemosis, eye pain	III	30	Partial	Yes	Yes
24	Tanriover et al.[2]	2007	34	F	Anisocoria, ptosis, headache, exotropia	III	20	Subtotal	Yes	No (improved)
25	Prabhu and Bruner[22]	2009	38	F	Headache, diplopia, ptosis, dizziness	III	35	Total	Yes	Yes
26	Goel and Shah[5]	2010	32	M	Headache, diplopia, ptosis	III	40	Subtotal	Yes	No (improved)
27			16	M	Headache, ptosis	III, IV, V	30	Total	Yes	Yes
28	Saetia et al.[23]	2011	41	M	Visual loss	II, III	45	Total	Yes	Yes
29	Present case	2013	37	F	Semicoma, anisocoria, acute hydrocephalus	III	50	Subtotal	Yes	Yes

F: female, M: male, ND: not described.

case, the PCA was the fetal type filled from the internal carotid artery, and the SCA was filled from the vertebral artery, so these vessels were shown in different views.[19] Our case illustrates the typical separation of the PCA and SCA. If the possibility of oculomotor schwannoma is high, the risk of complete oculomotor nerve palsy after surgery may be high. We emphasize the importance of detecting any separation of the SCA and PCA in cases of parasellar large tumors with third nerve palsy.

Conflicts of Interest Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this article. All authors who are members of the Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

References

1) Kovacs W: Ueber ein solitares Neurinom des Nervus oculomotorius. *Zentralbl Allg Pathol* 40: 518–522, 1927 (German)
2) Tanriover N, Kemerdere R, Kafadar AM, Muhammedrezai S, Akar Z: Oculomotor nerve schwannoma located in the

oculomotor cistern. *Surg Neurol* 67: 83–88; discussion 88, 2007
3) Asaoka K, Sawamura Y, Murai H, Satoh M: Schwannoma of the oculomotor nerve: a case report with consideration of the surgical treatment. *Neurosurgery* 45: 630–633; discussion 633–634, 1999
4) Broggi G, Franzini A: Value of serial stereotactic biopsies and impedance monitoring in the treatment of deep brain tumours. *J Neurol Neurosurg Psychiatr* 44: 397–401, 1981
5) Goel A, Shah A: "Interdural" oculomotor neurinoma: report of two surgically treated cases. *Acta Neurochir (Wien)* 152: 1721–1724, 2010
6) Hatakeyama H, Saito K, Nagatani T, Patel SJ, Yoshida J: Schwannoma in the crural cistern removed without permanent functional deficits—case report. *Neurol Med Chir (Tokyo)* 43: 95–99, 2003
7) Hiscott P, Symon L: An unusual presentation of neurofibroma of the oculomotor nerve. Case report. *J Neurosurg* 56: 854–856, 1982
8) Kachhara R, Nair S, Radhakrishnan VV: Oculomotor nerve neurinoma: report of two cases. *Acta Neurochir (Wien)* 140: 1147–1151, 1998
9) Kadota T, Miyawaki Y, Nakagawa H, Ishiguro S, Kuroda C: MR imaging of oculomotor nerve neurilemmoma. *Magn Reson Imaging* 11: 1071–1075, 1993
10) Kansu T, Ozcan OE, Ozdirim E, Onol B, Gurcay O: Neurinoma of the oculomotor nerve. Case report. *J Clin Neuroophthalmol* 2: 271–272, 1982
11) Katoh M, Kawamoto T, Ohnishi K, Sawamura Y, Abe H: Asymptomatic schwannoma of the oculomotor nerve: case report. *J Clin Neurosci* 7: 458–460, 2000
12) Katsumata Y, Maehara T, Noda M, Shirouzu I: Neurinoma of the oculomotor nerve: CT and MR features. *J Comput Assist Tomogr* 14: 658–661, 1990
13) Kurokawa Y, Uede T, Honda O, Honmou O: Successful removal of intracavernous neurinoma originating from the oculomotor nerve—case report. *Neurol Med Chir (Tokyo)* 32: 225–228, 1992
14) Leunda G, Vaquero J, Cabezudo J, Garcia-Uria J, Bravo G: Schwannoma of the oculomotor nerves. Report of four cases. *J Neurosurg* 57: 563–565, 1982
15) Lunardi P, Rocchi G, Rizzo A, Missori P: Neurinoma of the oculomotor nerve. *Clin Neurol Neurosurg* 92: 333–335, 1990
16) Mariniello G, Horvat A, Dolenc VV: En bloc resection of an intracavernous oculomotor nerve schwannoma and grafting of the oculomotor nerve with sural nerve. Case report and review of the literature. *J Neurosurg* 91: 1045–1049, 1999
17) Mehta VS, Singh RV, Misra NK, Choudhary C: Schwannoma of the oculomotor nerve. *Br J Neurosurg* 4: 69–72, 1990
18) Netuka D, Benes V: Oculomotor nerve schwannoma. *Br J Neurosurg* 17: 168–173, 2003
19) Niazi W, Boggan JE: Schwannoma of extraocular nerves: survey of literature and case report of an isolated third nerve schwannoma. *Skull Base Surg* 4: 219–226, 1994
20) Ohata K, Takami T, Goto T, Ishibashi K: Schwannoma of the oculomotor nerve. *Neurol India* 54: 437–439, 2006
21) Okamoto S, Handa H, Yamashita J: Neurinoma of the oculomotor nerve. *Surg Neurol* 24: 275–278, 1985
22) Prabhu SS, Bruner JM: Large oculomotor schwannoma presenting as a parasellar mass: A case report and literature review. *Surg Neurol Int* 1: 15, 2010
23) Saetia K, Larbcharoensub N, Wetchagama N: Oculomotor nerve schwannoma: a case report and review of the literature. *J Med Assoc Thai* 94: 1002–1007, 2011
24) Sarma S, Sekhar LN, Schessel DA: Nonvestibular schwannomas of the brain: a 7-year experience. *Neurosurgery* 50: 437–448; discussion 438–439, 2002
25) Schubiger O, Valavanis A, Hayek J, Dabir K: Neuroma of the cavernous sinus. *Surg Neurol* 13: 313–316, 1980
26) Schultheiss R, Kristof R, Schramm J: Complete removal of an oculomotor nerve neurinoma without permanent functional deficit. Case report. *Ger J Ophthalmol* 2: 228–233, 1993
27) Takano S, Endo M, Miyasaka Y, Yada K, Ohwada T, Takagi H: Neurinoma of the oculomotor nerve—case report. *Neurol Med Chir (Tokyo)* 30: 132–136, 1990

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