Surgical Treatment of Trochlear Nerve Schwannomas: Case Series and Systematic Review

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■ BACKGROUND AND OBJECTIVE: **Cranial nerve schwannomas almost always arise from sensory or mixed nerves. Motor cranial nerves, such as the trochlear nerve, are rarely associated with schwannomas. No consensus has yet been made for surgical intervention because of the low number of reported cases of trochlear nerve schwannomas. This study comprises a systematic review of the literature and our experience for surgically treated trochlear nerve schwannomas.**

■ METHODS: **Three databases (Web of Science, PubMed, and Cochrane Library) were searched without date restrictions. Studies were included if they were published in the English literature and presented patients of any age who underwent surgical treatment for trochlear schwannoma. Data extracted from the included studies were combined with our experience.**

■ RESULTS: **Forty-one studies, presenting 43 patients, met the inclusion criteria. The total number of patients was 45 after our experience was added. The most common symptoms were diplopia (62.2%), headache (46.7%), and motor weakness (37.8%). Mean age during the diagnosis was 45.1 years. Although the subtemporal transtentorial approach ($n = 14$) is the most preferred method, its application has decreased in recent years. In the last decade, the lateral suboccipital approach ($n = 11$) has gained popularity. Residual postoperative trochlear nerve deficit was detected in 81% of patients. The probability of neurologic deficit was not statistically associated with tumor volume ($P = 0.914$), location ($P = 0.669$), or resection rate ($P = 0.554$).**

■ CONCLUSIONS: **Although trochlear schwannomas are rare and their treatment involves challenges, total resection with the proper approach provides the most desirable results.**

INTRODUCTION

A schwannoma is a slow-growing benign nerve sheath tumor that can arise from both peripheral and cranial nerves.[1-3] Mostly, nerves that contain sensory fibers or mixed sensory and motor fibers are affected, when a schwannoma arises from a cranial nerve.[4] A pure sensory cranial nerve, the vestibulocochlear nerve, is affected by most cranial nerve schwannomas, followed by trigeminal and facial nerves.[1,4] Schwannomas rarely arise from pure motor cranial nerves, such as the trochlear nerve. Only 43 pathologically confirmed trochlear nerve schwannomas have been reported in the English literature.[5-45] In this study, we aimed to systematically review the reported cases in the literature and contribute with new cases of patients who were operated on because of trochlear schwannoma.

METHODS

This study was conducted following the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines. Three databases (Web of Science, PubMed, and Cochrane Library) were searched on June 1, 2021 without date restrictions using the algorithm ("trochlear nerve" or "fourth

Key words
- Nerve sheath tumor
- Neurilemmoma
- Neurinoma
- Schwannoma
- Surgery
- Trochlear nerve

Abbreviations and Acronyms
MR: Magnetic resonance
NF2: Neurofibromatosis type 2
SRS: Stereotactic radiosurgery

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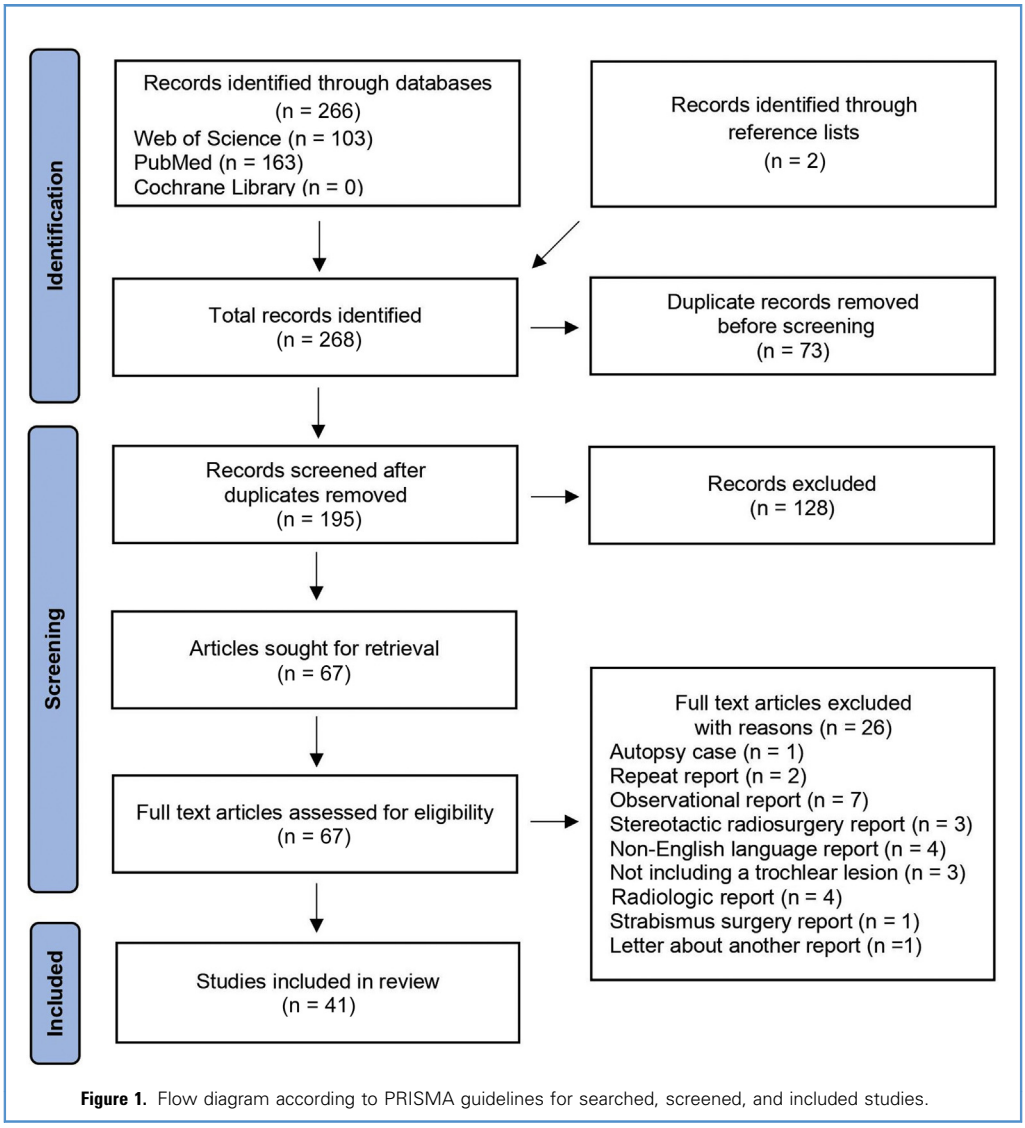

Figure 1. Flow diagram according to PRISMA guidelines for searched, screened, and included studies.

cranial nerve") and ("schwannoma" or "nerve sheath tumor" or"neurilemmoma" or "neurinoma"). The studies and relevant cases were included in the review if they 1) were published in English; 2) included patients of any age, and 3) included pathologically confirmed diagnosis of trochlear schwannoma via surgical treatment. Forty-one studies, including 43 cases in total, were identified in this way. Demographic, clinical, radiologic, surgical, and outcome data of all cases were extracted and summarized. After that process, our experience with a case series of 3 patients, one of which had been previously reported, was integrated into these data. For statistical analysis, we used the free Social Science Statistics software available at https://www.socscistatistics.com/ (accessed on June 1, 2021). A Fisher exact test was used to compare categorial variables. A P value <0.05 was considered statistically significant.

RESULTS

The results of the literature review are presented in a flowchart (Figure 1) designed according to the PRISMA guidelines. Of 268 records extracted from the databases and reference lists, after duplicates were removed and unrelated records excluded, 67 full-text articles were assessed for eligibility. Twenty-six studies were excluded with reasons. Forty-one studies were included in the review. There were 11 articles from the United States[5,6,12,13,25,30,33,37,39,41,44]; 8 articles from Japan[8,10,15,18,21,27,29,34]; 3 articles from Italy[14,31,40] and England[11,16,24]; 2 articles from Australia,[9,19] India,[20,26] Turkey,[23,43] Germany,[28,32] and Iran[36,42]; and 1 article from Spain,[7] France,[17] Israel,[22] China,[38] and Taiwan.[45] By combining these studies and our experience, the data of 45 patients in total were obtained. Information about the data of all cases included in the review is presented in Table 1.

Table 1. Table That Summarizing the Demographic, Clinical, Radiological, Surgical Approach and Follow-up Data of the Patients Included in the Review

Case Number	Reference	Age and Gender	Signs and Symptoms	Duration of Symptoms	Location	Size (mm)	Approach	Resection	Follow-Up	Outcome
1	King, 1976[5]	55, F	Headache Hemihypoesthesia Facial palsy (VII) Facial numbness (V) Decreased corneal reflex (V) Ataxia	3 months	Cisternal	30 × 40	Subtemporal transtentorial	Total	18 months	Trochlear nerve palsy Monoparesis (minimal, right leg)
2	Boggan et al., 1979[6]	32, F	Headache Diplopia (IV palsy) Facial palsy left Facial numbness, anisocoria Depressed gag reflex Hemiparesis, ataxia	2 years	Cisternal	40	Subtemporal transtentorial	Total	8 months	Trochlear nerve palsy Absent coronal reflex
3	Leunda et al., 1982[7]	54, F	Headache Diplopia	7 months	Cisternal	35	Subtemporal transtentorial	Total	6 month	Trochlear nerve palsy
4		16, M	Headache Diplopia	5 months	Cisternocavernous	45	Subtemporal transtentorial	Total	12 month	Trochlear nerve palsy
5	Yamamoto et al., 1987[8]	37, F	Diplopia Headache Nausea and vomiting,	3 months	Cisternal	10 × 9 × 7	Subtemporal transtentorial	Total	5 years	Trochlear nerve palsy
6	Garen et al., 1987[9]	18, F	Diplopia Ptosis Anisocoria	6 months	Cisternal	25	Subtemporal transtentorial	Total	ND	Trochlear nerve palsy
7	Tokuriki et al., 1988[10]	43, M	Hemiparesis Hemihypoesthesia Facial numbness Ataxia	13 months	Cisternal	ND	Subtemporal transtentorial	Subtotal	1 month	Trochlear nerve palsy
8	Maurice-Williams, 1989[11]	56, M	Diplopia, headache Hemiparesis Dysarthria Facial palsy	5 months	Cisternal	20	Lateral suboccipital approach	Total	2 years	Trochlear nerve palsy Slight spasticity
9	Samii et al., 1989[12]	53, F	Diplopia (III and IV palsy) Hemiparesis	ND	Cisternal	ND	Pterional	Total	ND	Trochlear nerve palsy
10	Gentry et al., 1991[13]	28, M	Headache Diplopia (IV palsy) V palsy, VII palsy	2 years	Cisternal (transition zone)	25 × 20 × 16	Suboccipital	Total	ND	Trochlear nerve palsy
11	Celli et al., 1992[14]	51, M	Headache, ataxia Diplopia (IV palsy) Hemiparesis	12 months	Cisternal	40 × 30	Subtemporal transtentorial	Total	5 years	Trochlear nerve palsy

12	Abe et al., 1994[15]	57, M	Headache Facial numbness	2 months	Cisternal	ND	Subtemporal transtentorial	Subtotal	4 months	Trochlear nerve palsy
13	Jackowski et al., 1994[16]	26, F	Headache Diplopia (IV palsy) Numbness on tongue (V) Papilledema, anisocoria (III) Altered memory	8 months	Cisternal	ND	Transtemporal transtentorial	Total	6 months	Trochlear nerve palsy
14	Dolenc and Coscia, 1996[17]	68, M	Diplopia Hemiparesis Hemianesthesia	2 years	Cisternal	30 × 25 × 25	Lateral suboccipital	Total	2 months	None
15	Beppu et al., 1997[18]	66, M	Tinnitus Hearing loss Vertigo Diplopia Facial (V2) hemianesthesia	5 years	Cisternal	2 × 2 × 1	Lateral suboccipital	Total	ND	Abductor nerve palsy
16	Santorenos et al., 1997[19]	35, F	Headache, Hemiparesis Bilateral bulbar paresis Emotional lability Reduced gag reflex	2 months	Cisternal	ND	Subtemporal transtentorial	Total	23 months	Trochlear nerve palsy
17	Nadkarni and Goel, 1999[20]	48, F	Inappropriate episodic laughter Depressed corneal reflex	6 months	Cisternal	ND	Subtemporal transtentorial	Total	6 months	Trochlear nerve palsy
18	Matsui et al., 2002[21]	61, M	Hemiparesis Hemianesthesia Diplopia (IV palsy)	2 months	Cisternal	ND	Presigmoid transpetrosal	Total	3 years	Trochlear nerve palsy
19	Veshchev and Spektor, 2002[22]	26, F	Atypical (burning) facial pain	3 months	Intratentorial	ND	Pterional	Total	4 months	Trochlear nerve palsy
20*	Ture et al., 2002[23]	31, M	Diplopia (IV palsy)	2 months	Cisternal	20 × 15	Infratentorial lateral subracerebellar	Total	ND	Trochlear nerve palsy
21	Shenouda et al., 2002[24]	49, M	Headache, ataxia Diplopia (III and IV palsy) Dysphagia Absent gag reflex Short-term memory loss Bilateral papilledema	3 weeks	Cisternocavernous	40 × 30	Presigmoid combined petrosal	Total	6 years	Trochlear nerve palsy Left-sided deafness
22	Du et al., 2003[25]	17, F	Headaches Diplopia Nystagmus Pitosis	3 months	Cisternocavernous	38 × 29 × 25	Orbitozygomatic pterional	Total	ND	None

F, female; M, male; ND, non-defined.
*This case is presented as case 1 in the review. In accordance with the time order, it is listed as the 20th case in the table.

Continues

Table 1. Continued

Case Number	Reference	Age and Gender	Signs and Symptoms	Duration of Symptoms	Location	Size (mm)	Approach	Resection	Follow-Up	Outcome
23	Shenoy and Raja, 2004[26]	54, F	Headache, ptosis Hemiparesia, hemianesthesia Facial palsy Dysarthria, Dysphagia Trochlear nerve palsy	2 months	Cisternal	ND	Subtemporal transtentorial	Total	1 year	Hemianesthesia Trochlear nerve palsy
24	Ohba et al., 2006[27]	42, M	Hemiparesis Oculomotor nerve palsy Trochlear nerve palsy Facial hypesthesia Facial palsy	6 weeks	Cisternal	30	Anterior transpetrosal	Total	4 months	Trochlear nerve palsy
25	Gerganov et al., 2007[28]	52, F	Diplopia, ataxia Facial palsy Hearing loss Hemiparesis	10 months	Cisternal	25 (solid component) + 29 (cystic component)	Retrosigmoid	Total	28 months	Trochlear nerve palsy Facial hypesthesia
26	Kohama et al., 2009[29]	47, F	Hemiparesis Ataxia	5 months	Cisternal	ND	Posterior transpetrosal	Total	ND	Trochlear nerve palsy
27	Elmalem et al., 2009[30]	ND	Trochlear nerve palsy	6.9 months	ND	20	ND	ND	ND	ND
28	Elmalem et al., 2009[30]	ND	Incidental (after trauma)	0	Cisternocavernous	50	ND	ND	ND	ND
29	Bartalena et al., 2010[31]	50, F	Diplopia	3 months	Cisternal	25	Subtemporal transtentorial	Total	15 months	Trochlear nerve palsy
30	Younes et al., 2012[32]	65, F	Dizziness, vertigo Diplopia (IV palsy) Monoparesis	ND	Cisternal	17	Pterional	Subtotal	2 years	None
31	Boucher and Michael, 2014[33]	64, M	Diplopia (IV palsy) Headache	2 months	Cisternal	9 × 7 × 11	Anterior transpetrosal	Total	6 months	Trochlear nerve palsy
32	Hatae et al., 2014[34]	44, M	Hiccups, trochlear nerve palsy	0.4 months	Cisternal	27 × 27 × 30	Anterior transpetrosal	Subtotal	1 year	None
33	Inoue et al., 2015[35]	52, M	Diplopia Hemiparesis Ataxia	2 years	Cisternal	ND	Subtemporal + retrosigmoid	Subtotal	15 months	Trochlear nerve palsy
34	Samadian et al., 2015[36]	63, M	Headache Diplopia	8 months	Cisternal	ND	Retrosigmoid	Total	ND	Trochlear nerve palsy
35	Chaudhry et al., 2016[37]	24, F	Headache	0.4 months	Cisternal (transional)	24 × 22 × 31	Paraoccipital transtentorial	Total	6 months	Trochlear nerve palsy

36	Liu et al., 2016[38]	65, M	Facial pain Diplopia	4 months	Cisternal	ND	Retrosigmoid	Total	2 years	Trochlear nerve palsy
37	Nesvick et al., 2017[39]	50, F	Trigeminal neuralgia	6 months	Cisternal	17 × 9 × 8	Retrosigmoid	Total	3 months	Intermittent diplopia
38	Cunha et al., 2017[40]	29, F	Headache, diplopia Blurred vision Nausea and vomiting	3 years	Cisternal (transitional)	ND	Infratentorial supracerebellar	Subtotal	ND	Trochlear nerve palsy
39	Torun et al., 2018[41]	24, M	Headache Diplopia Ataxia	3 years	Cisternocavernous	35 × 20 × 15	Subfrontal pterional	Total	2 year	Trochlear nerve palsy
40	Farrokhi et al., 2018[42]	12, M	Headache Ataxia Diplopia	2 months	Cisternal (transitional)	20	Suboccipital Transvelar	Total	12 years	None
41	Erdem et al., 2019[43]	49, F	Hemihypoesthesia	2 months	Cisternal	ND	Retrosigmoid	Total	2 months	None
42	Tolisano et al., 2019[44]	41, M	Headache Ataxia	6 months	Cisternal	18 × 26	Transpetrosal retrolabyrinthine	Total	ND	Trochlear nerve palsy
43	Lan et al., 2020[45]	65, M	Headache Dizziness	0	Cisternal (transitional)	ND	Infratentorial supracerebellar	Subtotal	ND	ND
44	Present study, 2021	41, M	Facial hemihypoesthesia (V) Left hemiparesis Decreased gag reflex	5 years	Cisternal	32 × 19 × 22	Lateral suboccipital supracerebellar transtentorial	Total	5 years	None
45		71, F	Diplopia (trochlear nerve palsy) Hemiparesis	5 months	Cisternocavernous	35 × 25 × 26	Lateral suboccipital supracerebellar transtentorial	Total	1 year	Trochlear nerve palsy

F, female; M, male; ND, non-defined.

*This case is presented as case 1 in the review. In accordance with the time order, it is listed as the 20th case in the table.

Table 2. Signs and Symptoms of the 45 Cases Included in the Review

Symptoms	n	%
Diplopia	28	62.2
Headache	22	48.9
Ataxia	13	28.9
Vertigo	6	13.3
Nausea and vomiting	3	6.7
Facial pain	4	8.9
Personality change	2	4.4
Memory deficits	2	4.4
Hiccups	1	2.2
Pathologic laughter	1	2.2
Dysphagia	1	2.2
Signs		
Long tract signs		
Motor deficits	17	37.8
Sensorial deficits	9	20
Hyperreflexia	6	13.3
Cranial nerve signs		
Third cranial nerve	8	17.8
Fourth cranial nerve	23	51.1
Fifth cranial nerve	10	22.2
Sixth cranial nerve	0	
Seventh cranial nerve	3	6.7
Eighth cranial nerve	2	4.4
Inferior cranial nerves	6	13.3
Cerebellar signs	9	20
Papilledema	3	6.7

The signs and symptoms of the cases included in this review are shown in Table 2. Double vision was the most common symptom. Twenty-eight patients (62.2%) described that they had diplopia before or during admission. Headache (n = 22; 46,7%) was also among the cardinal symptoms. The third most common symptom was ataxia (n = 13; 28.9%). The most common finding (n = 23; 51.1%) in cranial nerve examinations was superior oblique muscle paralysis, which indicates the trochlear cranial nerve disorder. This finding was followed by others pointing to trigeminal nerve (n = 10; 22.2%) and oculomotor nerve (n = 8; 17.8%) disorders. Long tract findings were found in 18 cases. Among the cases that showed these findings, motor deficit (n = 17; 37.8%) was detected in almost all. Sensory deficits were reported in 9 cases (20%) and hyperreflexia in 6 cases (13.3%). In cerebellar tests, pathognomonic findings were reported in 9 cases (20%).

The demographic, radiologic, and surgical characteristics of 45 cases included in the study are shown in Table 3. Mean age during the diagnosis was 45.1 ± 15.6 years (range, 16–71 years). The age of diagnosis peaked in the 51–55 years age range. The female/male ratio was 1:1.05. The mean time from the appearance of symptoms to diagnosis was 10.1± 14.4 months. In most cases (n = 37; 84%) included in the review, tumors localized in cisterns. Tumors were cisternocavernous in 6 cases and intertentorial in 1 case.

Table 3. Characteristics of Cases Included in the Review

Mean age (years) (n = 43)*	45.1 ± 15.6
Gender (n = 43)*	
Female	21
Male	22
Duration of symptoms (months)	10.1 ± 14.4
Tumor location (n = 44)*	
Cisternal	37
Quadrigeminal cistern (transitional)	5
Ambient and crural cisterns	32
Cisternocavernous	6
Intratentorial	1
Maximum diameter (mm) (n = 30)*	27.9 ± 11. 6
Approaches (n = 43)*	
Subtemporal transtentorial	14
Lateral suboccipital	11
Transpetrosal	7
Pterional	5
Infratentorial supracerebellar	2
Medial suboccipital	2
Supratentorial interoccipital	1
Subtemporal + retrosigmoid combined	1
Resection rate (n = 43)*	
Total	36
Subtotal	7
Outcome (n = 42)*	
Cranial nerve deficits	35
Fourth nerve	34
Fifth nerve	2
Sixth nerve	1
Eighth nerve	1
Long tract deficits	3
Monoparesis	1
Spasticity	1
Hemianesthesia	1
Without any deficit	7

*The relevant results are presented in the number of cases indicated.

Measurement data on the size of the tumor were available in 30 cases. Mean tumor size was observed as 27.9 ± 11.6 mm.

Surgical approach choice was reported in 43 cases. Supratentorial approaches were used in 27 cases, infratentorial approaches in 15 cases, and a combined approach in 1 case. A subtemporal transtentorial approach ($n = 14$) is the most performed approach among supratentorial approaches. Lateral suboccipital approaches ($n = 11$) constituted most infratentorial approaches. The suptemporal approach was preferred in 12 of the 17 cases reported before 2000. In 2 of 10 cases reported between 2001 and 2010, the subtemporal approach was preferred. In 16 cases reported in 2011 and after, the suptemporal approach was not performed. At the same year interval, lateral suboccipital approaches were performed in 3 cases (2000 and before), 2 cases (2001–2010), and 6 cases (2011 and after).

For the all cases included in the review, the mean follow-up time was 22 ± 28.6 months. Postoperative outcome information was provided in 42 cases, 7 of which had no residual neurologic deficit. Also, in 35 cases, there were various neurologic deficits at the end of the follow-up period. The most common of these deficits was trochlear nerve palsy, which was observed in 34 cases. To investigate the factors on residual neurologic deficit, 2 groups of cases were formed, with deficits ($n = 35$, group A) and without deficits ($n = 7$, group B). The mean tumor measurements for groups A and B were 27.3 ± 11.6 and 27.8 ± 7.9, respectively. There was no statistical difference between the groups ($P = 0.914$). In group A, total resection was achieved in 31 cases and subtotal resection in 4 cases, and in 5 cases and 2 cases in group B. There was no statistical difference ($P = 0.554$) between the groups in terms of resection rate (total or subtotal). Regarding tumor localizations, in group A, 30 cases were cisternal, 4 cases were cisternocavernous, and 1 case was intratentorial. Also, in group B, 5 cases were cisternal, and 1 case was cisternocavernous. Tumor localizations were not different between the groups ($P = 0.669$).

Case 1

Herein, we present previously reported case and follow-up data.[23] A 31-year-old man, who reported intermittent diplopia for 2 months, was admitted to our department. Neurologic examination showed left trochlear nerve palsy. Magnetic resonance (MR) images showed a well-circumscribed mass lesion (20 × 15 mm) located in the left ambient cistern and compressing the pons and mesencephalon (Figure 2). Left-sided global slowing was observed at brainstem auditory evoked potential latencies.

A left-sided infratentorial lateral supracerebellar approach was performed via retromastoid craniotomy in a lateral decubitus position. A yellow soft well-circumscribed mass localized inside the ambient cistern, which was not cleavable from the trochlear nerve, was completely removed via dissection from the surrounding structures.

No additional neurologic incident was observed during the postoperative course. A schwannoma, consisting of diffuse Schwann cells with mild cellular atypia and with neither mitosis nor necrosis, was shown on histopathologic examination. Postoperative MR imaging verified total removal. During the follow-up, the right trochlear nerve palsy persisted and there was no recurrence. Also, trochlear nerve palsy was asymptomatic and detected only on neurologic examination. Neither daily life modification nor strabismus surgery was needed.

Case 2

A 41-year-old female patient with a history of operation and consequent stereotactic radiosurgery (SRS) 5 years previously was referred to our department after the tumor was observed to have progressed at follow-up. Neurologic examination showed hypoesthesia in the frontal and maxillary branches of the trigeminal nerve, decreased gag reflex, and left hemiparesis (4/5).

MR imaging showed a peripherally contrast-enhanced mass lesion (32 × 19 × 22 mm) that was located in the right crural, prepontine, and interpeduncular cisterns. Mass lesion was shown, containing solid and cystic components, compressing the pons, mesencephalon, and crus cerebri, and also displacing the basilar artery (Figure 3).

The mass was totally removed via a lateral supracerebellar transtentorial approach via lateral suboccipital craniotomy in the semilateral position. Initially, an internal decompression was performed during tumor removal. The tumor was adjacent to the third, fifth, and sixth nerves and extended to the point where the fourth nerve entered the cavernous sinus. Meticulous dissection of the arachnoid plane around the tumor enabled a removal that ensured preservation of surrounding eloquent structures.

Total removal of the lesion was verified via postoperative MR imaging (Figure 3). Histopathologic results showed that the lesion comprised schwannoma. Histopathologic examinations showed that the pathologic result of the lesion was schwannoma. No new neurologic deficit was detected after the operation and the previous deficits resolved in the postoperative period. No recurrence was detected during 3 years of follow-up.

Case 3

A 71-year-old woman, presenting with diplopia for 5 months and left hemiparesis for 3 months, was referred to our department. Neurologic examination showed left trochlear nerve palsy and left hemiparesis that was more noticeable in the lower extremity (3/5) compared with the upper extremity (4/5).

MR imaging showed a well-circumscribed heterogeneously contrasted mass lesion (35 × 25 × 26 mm) located in the right half of the pons extending toward the prepontine cistern and posterior part of the cavernous sinus. Mass lesion was compressing the pons, mesencephalon, and aqueductus sylvius and was also displacing the basilar artery, posterior cerebral artery, posterior communicating artery, and third cranial nerve (Figure 4).

A lateral supracerebellar transtentorial approach via lateral suboccipital craniotomy in the semilateral position was performed and the mass was totally excised. The mass, which was adjacent to the brainstem and invaded the cavernous sinus, was observed to originate from the fourth nerve and could not be separated from the nerve (see Surgical Video 1).

Video available at www.sciencedirect.com

Intraoperative MR imaging verified total removal. Minimal contrast enhancement at the tentorial edge within the cavernous sinus was noticed in the MR imaging 3 months after operation (Figure 4). No relapse was detected during 1 year of follow-up. Histopathologic evaluation showed that the lesion comprised schwannoma. In the postoperative period, left

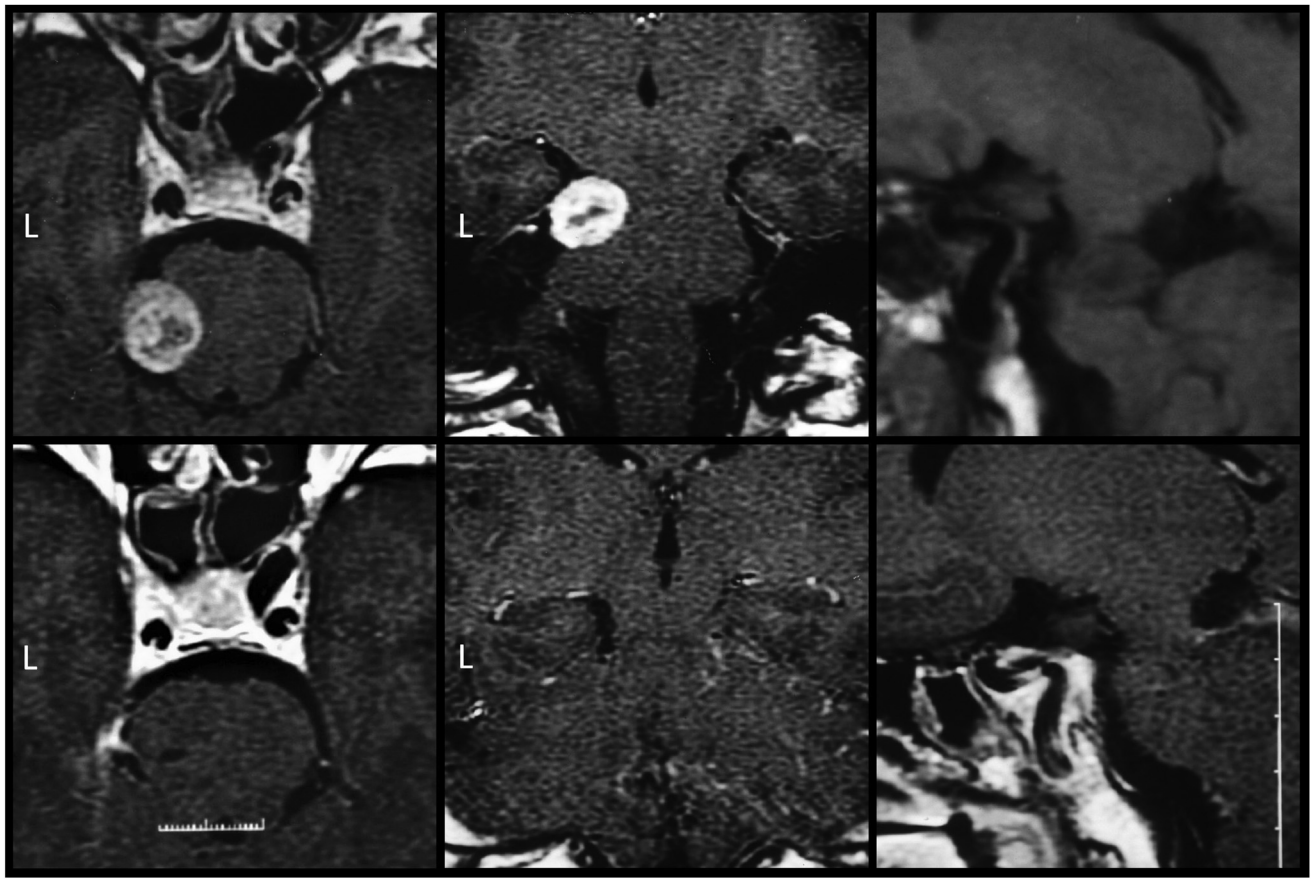

Figure 2. (*Upper row*) Before the operation, T1-weighted contrast-enhanced axial (*left*), C+ coronal (*middle*), and sagittal (*right*) section magnetic resonance images (MRIs) showing a well-demarcated cisternal-type trochlear schwannoma, which was located within the ambient cistern at the edge of the tentorial incisura. (*Lower row*) After the operation, T1-weighted contrast-enhanced axial (*left*), contrast-enhanced coronal (*middle*), and sagittal (*right*) section magnetic resonance images showing that the mass was totally removed.

hemiparesis resolved but left trochlear nerve palsy persisted. Mild visual symptoms were reported during her daily routine and minor positional changes were sufficient for proper vision.

DISCUSSION

Trochlear schwannomas are rare tumors. Since the first reported case[5] in 1976, 45 patients have been surgically treated for this tumor. When viewed in 5-year intervals, the number of reported cases increased from 2 to 9 from 1976–1981 to 2016–2021. Several factors have possibly caused the increase in the number of reported cases of trochlear schwannomas. The increased access to diagnostic radiologic tests such as MR imaging is probably the most important factor.[46] Another factor may be the increased average life expectancy, especially in developed countries.[47] In addition, another factor may be increased rates of medical admissions as a result of symptoms such as diplopia.

According to the results of this review, cardinal symptoms of patients who were referred for surgical treatment for trochlear schwannoma include double vision, headache, and ataxia. In addition, many atypical causes of admission have been reported. If a triad of trochlear schwannoma were to be defined, it would likely include: 1) diplopia (trochlear nerve palsy), 2) long tract signs, and 3) headache. However atypical presenting symptoms such as burning facial pain,[22] hiccups,[34] and pathologic laughter[20] have been reported in some cases.

Intracranial schwannomas can occur at any age, although they are extremely rare in childhood.[1] The highest incidence is detected in the third, fourth, and sixth decades of life.[1] According to a national cohort of vestibular schwannomas from Denmark, the median age at diagnosis was 49 years in 1976, which increased to 60 years in 2015.[48] In this review, similar results were obtained for trochlear schwannomas. We found that the median age was 49 years in the recent review, with the highest incidence in the fifth and six6th decades of life. Intracranial schwannomas usually show no difference in occurrence between females and males.[1] In the national vestibular schwannoma cohort from Denmark with 3637 patients, the female/male ratio was close to 1, with 1804 females and 1833 males.[48] In our study, a similar result was obtained, with a female/male ratio of 0.95.

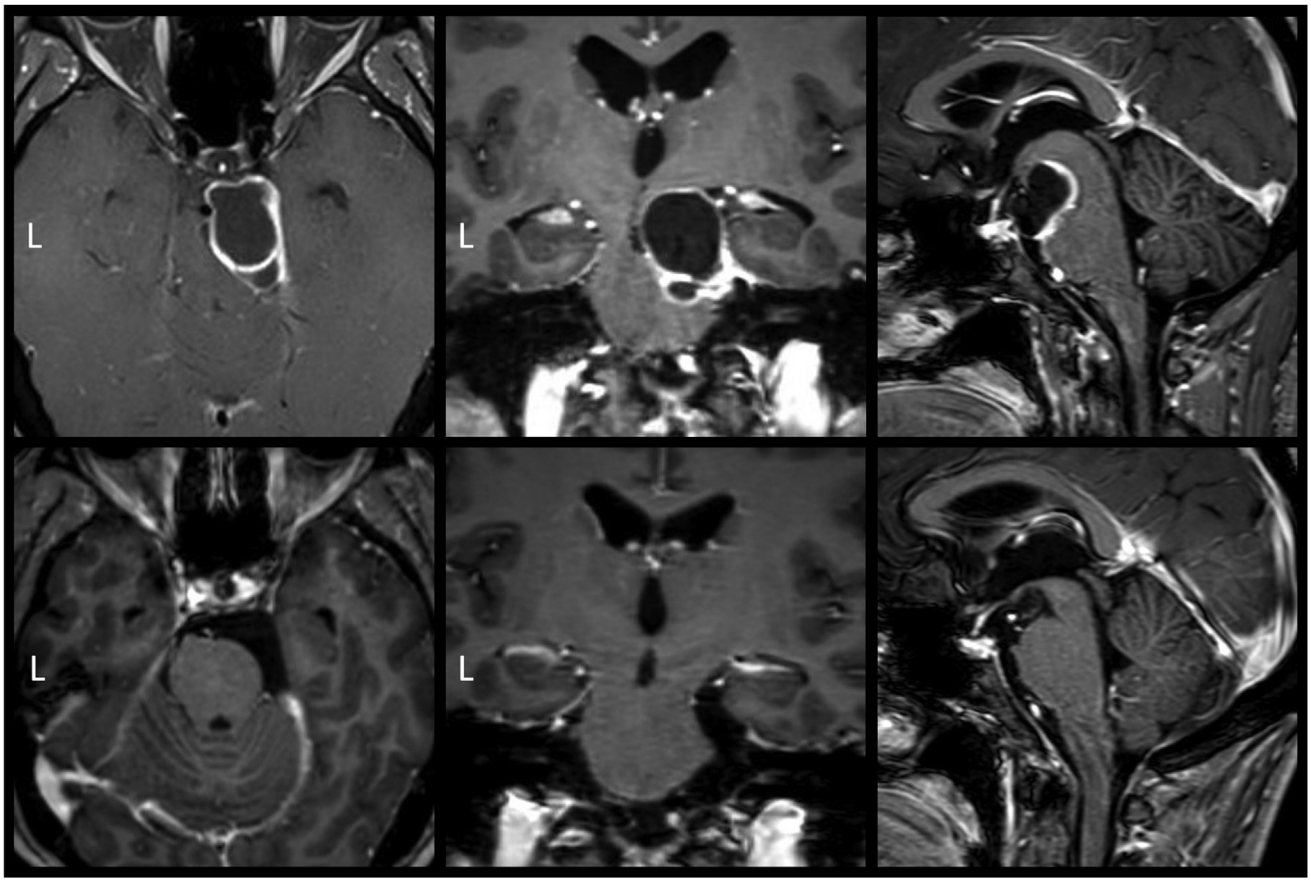

Figure 3. (*Upper row*) Before the operation, T1-weighted contrast-enhanced axial (*left*), contrast-enhanced coronal (*middle*), and contrast-enhanced sagittal (*right*) section magnetic resonance images showing a peripherally contrast-enhanced trochlear schwannoma located within the right crural, prepontine and interpeduncular cisterns, which was compressing the pons, mesencephalon, and crus cerebri. (*Lower row*) After the operation, T1-weighted contrast-enhanced axial (*left*), contrast-enhanced coronal (*middle*), and contrast-enhanced sagittal (*right*) section magnetic resonance images showing that the mass was totally removed.

Trochlear schwannomas mostly present as isolated intracranial mass lesions. However, their association with another intracranial space-occupying lesion has occasionally been encountered.[15,18] A case of trochlear nerve schwannoma associated with a giant thrombosed vertebral artery aneurysm,[15] and another case of trochlear nerve schwannoma accompanied by a cerebellopontine angle meningioma,[18] have been reported.

Typically trochlear schwannomas can be classified into 3 types according to the tumor location: 1) cisternal, 2) cisternocavernous, and 3) cavernous types.[14] Cisternal-type trochlear schwannomas were observed in most of the reported cases. Cisternocavernous and cavernous types were relatively less diagnosed. In our series, 2 tumors (cases 1 and 2) were the cisternal type, whereas the other tumor (case 3) was compatible with the cisternocavernous type.

Supratentorial or infratentorial median approaches were preferred for tumors located in the ambient cistern. In 1 case, the supratentorial paraoccipital transtentorial approach was preferred, whereas infratentorial approaches such as infratentorial supracerebellar and suboccipital transvelar approaches were performed in another 4 cases. In all cases except for these 5, lateral approaches were preferred. The subtemporal approach is the most frequently preferred approach, followed by the lateral suboccipital approach. When all cases in this review were evaluated without including tumors located in the ambient cistern, the rate of choosing the subtemporal approach was 75% in cases reported before 2000. The same ratio is 20% and 0%, respectively, in the periods 2001–2010 and 2011–present, whereas, these rates for the lateral suboccipital approach were 18.7%, 20%, and 50%, respectively. Especially in the last decade, the subtemporal approach has lost its popularity, whereas the lateral suboccipital approaches have become more widely used. This change may have several causes. The first possible cause is complications related to the subtemporal approach. Probable problems, such as temporal lobe contusion or injury to the vein of Labbé, are associated with temporal lobe elevation, which is required during the subtemporal approach.[49]Another plausible cause is that larger portions of

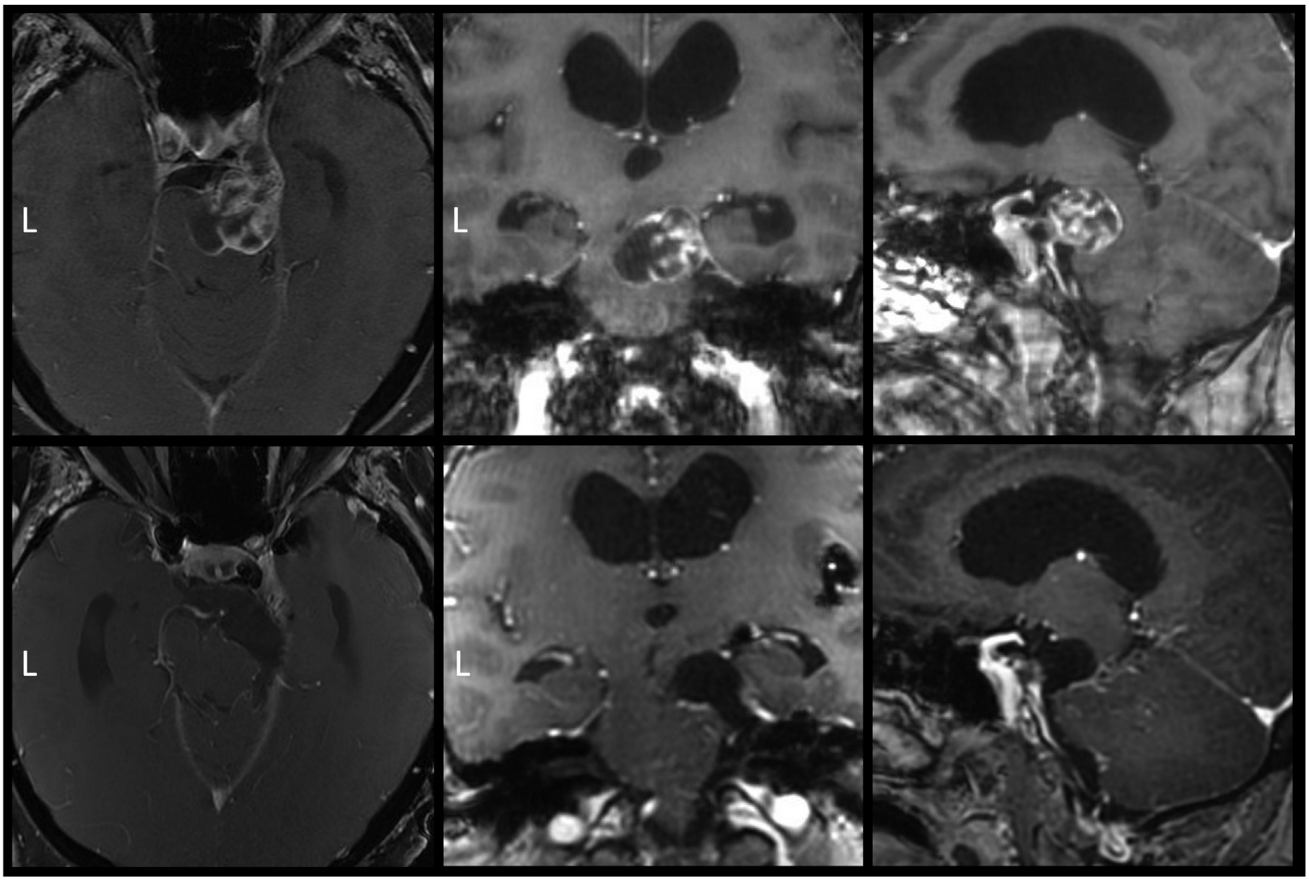

Figure 4. (*Upper row*) Before the operation, T1-weighted contrast-enhanced axial (*left*), contrast-enhanced coronal (*middle*), and contrast-enhanced sagittal (*right*) section magnetic resonance images showing a well-circumscribed heterogeneously contrasted trochlear schwannoma located in the right half of the pons extending toward the prepontine cistern and posterior part of the cavernous sinus, which was compressing the pons, mesencephalon, and aqueductus sylvius and also displacing the basilar artery. (*Lower row*) After the operation, T1-weighted contrast-enhanced axial (*left*), contrast-enhanced coronal (*middle*), and contrast-enhanced sagittal (*right*) section magnetic resonance images are presented.

trochlear schwannomas are infratentorial.[23] Therefore, it is more convenient to approach using an infratentorial route.[23]

A lateral supracerebellar transtentorial approach via lateral suboccipital craniotomy was performed in the 3 cases that present our experience. One of the main factors in choosing this approach was to avoid complications associated with the subtemporal approach, such as temporal lobe contusion and injury to the vein of Labbé. Early cerebrospinal fluid drainage, after suboccipital craniotomy, is possible with this approach. Cerebrospinal fluid drainage through cisterna magna dissection provides cerebellar relaxation, which enables an appropriate surgical passage during the lateral supracerebellar approach. The transtentorial route ensures intervention to the infratentorial and supratentorial parts of trochlear schwannomas. Also, with this approach, it is possible to remove the part of the tumor that extends into the cavernous sinus, if present.

The postoperative follow-up results of the review showed that fourth nerve palsy occurred in 81% of cases. The main reason for this situation may be that the schwannoma originates within the trochlear nerve in most cases and that separating the tumor from the fourth nerve is unattainable during surgery.[23] Younes et al.[32] suggested subtotal tumor excision for preserving the trochlear nerve. In their case, they used the pterional transsylvian approach and resected subtotally the tumor located in the ambient cistern. They left a remnant of tumor capsule to preserve fourth nerve function. Consequently, these investigators reported that the patient was free of diplopia with a stable tumor remnant on MR imaging after 2 years of follow-up. Contrarily, in the results of our review, when all cases were included, no better outcome was achieved with subtotal or total resection. Also, during the follow-up, rapid enlargement[44] of trochlear schwannomas and presenting with intratumoral hemorrhage[27,34,38] have been reported. In addition, most patients tolerate fourth nerve palsy well, and strabismus surgeries are available.[50,51] Considering all these points, performing a total resection, if possible, is favorable even if the fourth nerve cannot be preserved.

Observation is a valid option in nonsymptomatic trochlear schwannomas.[41] Sequential MR imaging with multidisciplinary follow-up was suggested in asymptomatic patients in the treatment algorithm proposed in the review by Torun et al., which screened 85 cases.[41] According to this review, no intervention was made in 15 patients and only 1 patient had clinical worsening during follow-up.[12]

SRS can be considered symptomatic (minor symptoms; likewise trochlear nerve palsy, and without brainstem compression, other cranial nerve palsies and long tract sings) small trochlear schwannomas.[41,52,53] According to a systematic review of 12 patients who presented with diplopia only,[41] after SRS, symptoms of 8 patients improved at a mean follow-up of 22 months. A study that evaluated the role of Gamma Knife SRS (Elekta, Stockholm, Sweden) in schwannomas[52] reported that the lesions decreased in 3 patients with trochlear schwannoma after a mean follow-up of 44 months. In a recent study reporting outcomes after SRS for non-vestibular schwannomas (11 of 23 trochlear schwannomas),[53] local tumor control rates were 96% at 1 year and 86% at 10 years.

Neurofibromatosis type 2 (NF2) is a rare genetic disorder associated with central nervous system tumors, caused by mutation in the NF2 gene on chromosome 22q12.[54] Vestibular schwannomas are the characteristic lesion, affecting 95% of individuals, and usually appear bilaterally.[54] Also, association of NF2 with trochlear schwannomas has been reported.[9] In a case series, NF2 was reported in 1 of 3 patients with trochlear schwannoma managed with Gamma Knife SRS.[52] In contrast, in another SRS series, none of 11 patients with trochlear schwannoma had NF2.[53] Another review that screened patients with pathologic diagnosis of trochlear schwannoma reported that none of the 33 patients had signs associated with NF2.[39]

Schwannomas composed of a mixture of compactly arranged Antoni type A cells and loosely arranged Antoni type B cells contain variable cellularity and water content.[55] Thus, trochlear schwannomas appear heterogeneously hyperintense on T2-weighted MR images.[55] The route of the nerve, interpeduncular, ambient, and prepontine cisterns is a typical localization of trochlear schwannomas. A slow growth pattern without any significant edema in surrounding neural tissue is an evidence for schwannomas on both MR and computed tomography images.[55] Some lesions can be confused with trochlear schwannomas. Specific features can be discussed for lesions in the differential diagnosis. Dural tail enhancement is a distinctive appearance for meningiomas.[56] Multiple nerve involvement and enhancement in meningeal surfaces are usually observed in leptomeningeal diseases.[57] Aneurysms can be distinguished with vascular flow void.[58]

CONCLUSIONS

Trochlear nerve schwannomas are rare tumors that require surgical intervention when presenting with signs of cranial nerves and long tracts compression. Although the treatment process includes challenges, satisfactory results can be obtained with appropriate surgical approaches.

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