CASE REPORT

Schwannoma of Extraocular Nerves: Survey of Literature and Case Report of an Isolated Third Nerve Schwannoma

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Abstract—An unusual case of schwannoma arising from the third cranial nerve in a thirteen year old male is reported. The patient presented with paresis of the right oculomotor nerve and ipsilateral hemiparesis. The clinical features of this case are discussed and the pertinent medical literature reviewed. (*Skull Base Surgery, 4(4):219–226, 1994*)

Schwannomas arising from an oculomotor nerve are exceedingly rare, particularly when there is no evidence of neurofibromatosis. The rarity of reported cases may, in part, be due to the difficulty in defining the point of origin for nerve sheath tumors arising in the parasellar region. The authors herein report a case of parasellar schwannoma that is felt to have originated from the third cranial nerve in a 13-year-old male.

CASE REPORT

A 13-year-old left-handed male was admitted to the pediatric neurology service at the University of California, Davis Medical Center on August 23, 1985. He had a 2-week history of progressive right-sided weakness that was associated with double vision on gaze to the left, slurring of his speech, and left occipitoparietal headache. During the 2 days before admission the patient's condition had worsened to where he was unable to lift his right arm or walk. His past medical history was remarkable for asthma and congenital hip dysplasia.

Physical examination demonstrated right-sided ptosis and slight anisocoria with the right pupil 4 mm and the left pupil 3 mm. Both pupils were briskly reactive to light. Adduction was decreased in the right eye. There was right central seventh nerve weakness, marked right hemiparesis worse in the leg than in the arm, and hyperreflexia. Babinski's sign was present on the right. No sensory deficits were found.

Computed tomography of the brain revealed a right parasellar, contrast-enhancing lesion that appeared to be partly cystic (Fig. 1). The lesion extended into the middle fossa and interpeduncular cistern, displacing the brain stem to the left against the tentorial edge. The patient was started on steroids and anticonvulsants. He underwent a cerebral angiogram that revealed stretching and displacement of the basilar, right posterior cerebral, superior cerebellar, and anterior inferior cerebellar arteries. The right superior cerebellar and anterior inferior cerebellar arteries were displaced posteroinferiorly, whereas the posterior cerebral, internal carotid, and proximal middle cerebral arteries were displaced superiorly. The lesion was relatively avascular (Fig. 2).

On hospital day 5 the tumor was explored through a right frontotemporal craniotomy. A rubbery mass with multiple cystic areas and loose yellowish tissue was found to arise from the posterolateral superior aspect of the cavernous sinus. It extended laterally, posteriorly, and inferiorly into the posterior and middle fossae. During the initial dissection, the internal carotid artery, its bifurcation, and the posterior communicating artery were demonstrated; however, the third cranial nerve could not be identified. The tumor was debulked using the carbon dioxide laser and standard microdissection. Continued dissection allowed tumor to be free from the lateral and anterior aspect of the brain stem and tentorium. This allowed the proximal third nerves to be identified. A film of tumor was left on the superiorlateral wall of the cavernous sinus that was felt to be somewhat full. Patholog-

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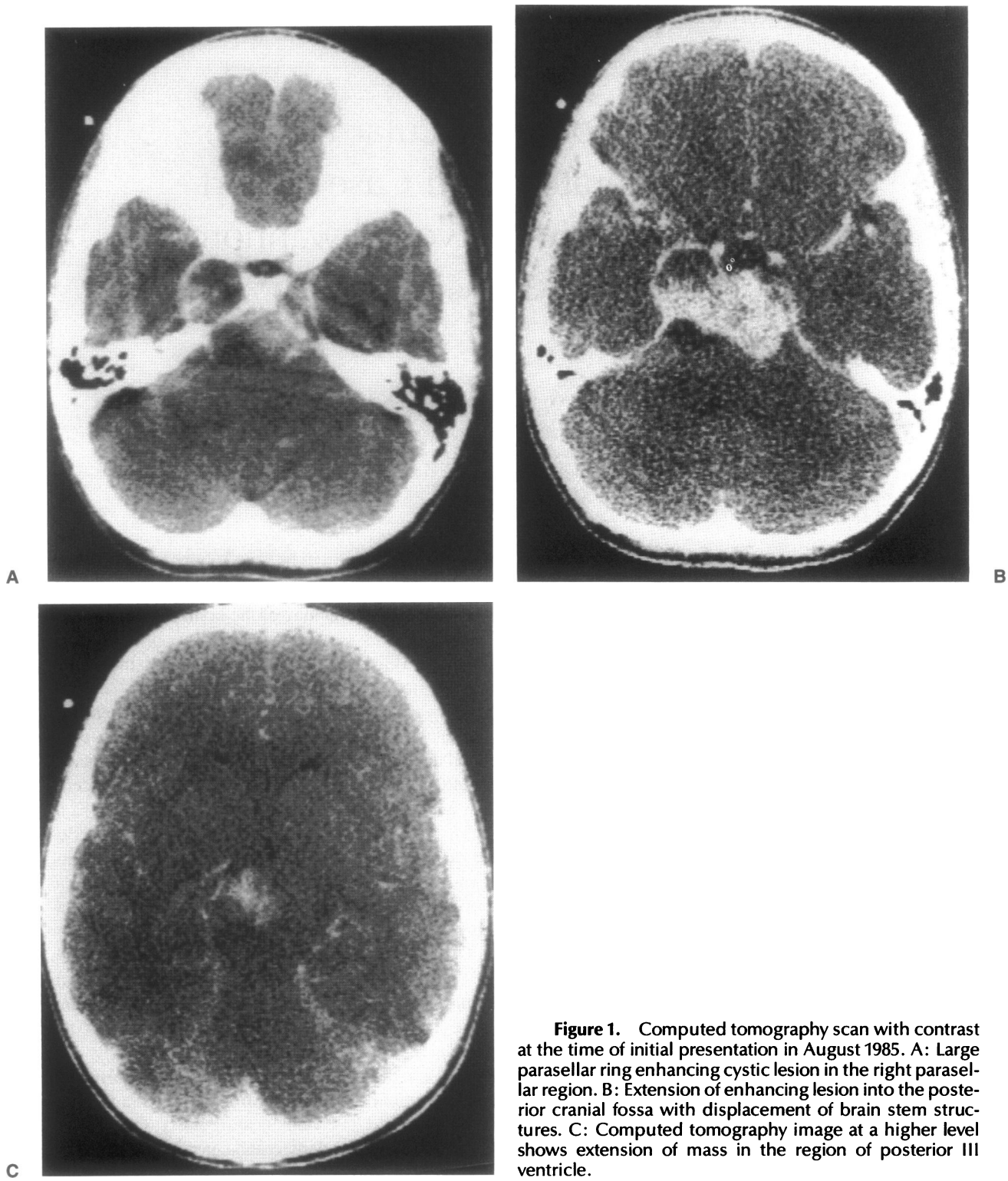

Figure 1. Computed tomography scan with contrast at the time of initial presentation in August 1985. A: Large parasellar ring enhancing cystic lesion in the right parasellar region. B: Extension of enhancing lesion into the posterior cranial fossa with displacement of brain stem structures. C: Computed tomography image at a higher level shows extension of mass in the region of posterior III ventricle.

ical examination revealed tumor histology compatible with a schwannoma (Fig. 3).

Postoperatively the patient's right hemiparesis resolved completely, but the right third nerve paresis persisted. The pupil reacted sluggishly to light. He complained of diplopia in all fields of gaze except the far right lateral gaze. Right fifth nerve function was normal. His immediate postoperative course was complicated by bronchospasm, fever, and hyponatremia, all of which resolved with medical management. The patient was discharged 2 weeks after the operation. On follow-up clinic visits his right third nerve function improved gradually. A repeat

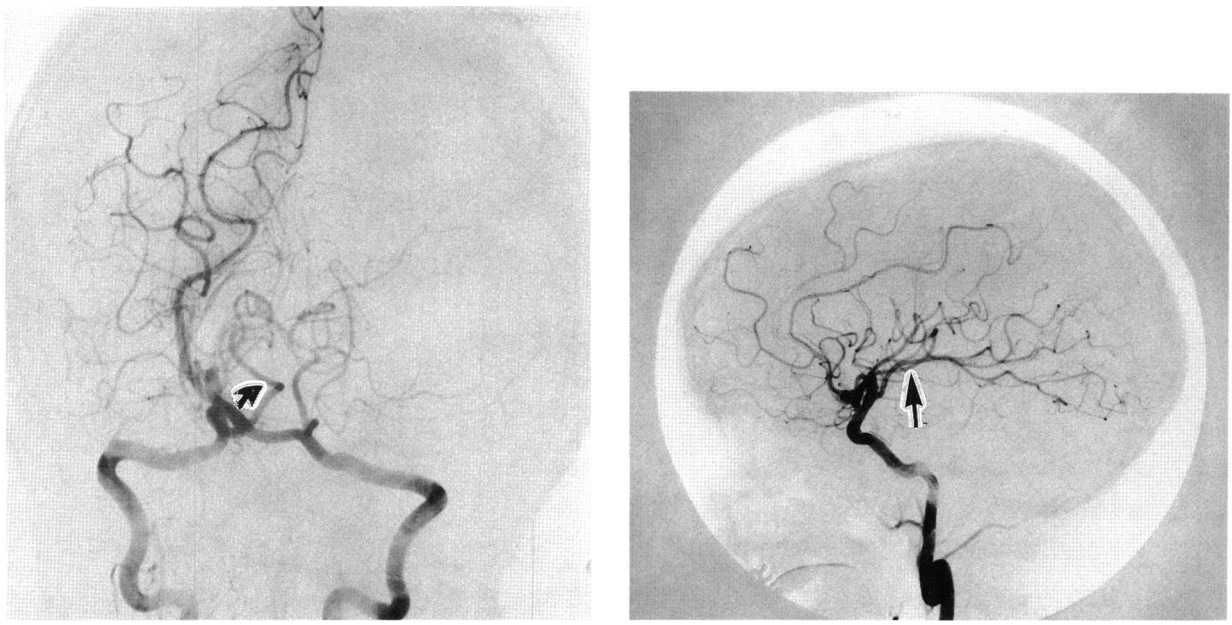

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Figure 2. Preoperative angiogram in 1985. A: Injection of posterior circulation: curved arrow indicates downward displacement of superior cerebellar artery by the mass lesion. B: Injection of anterior circulation: arrow indicates superior displacement of the posterior cerebral artery, which appears to arise from internal carotid artery.

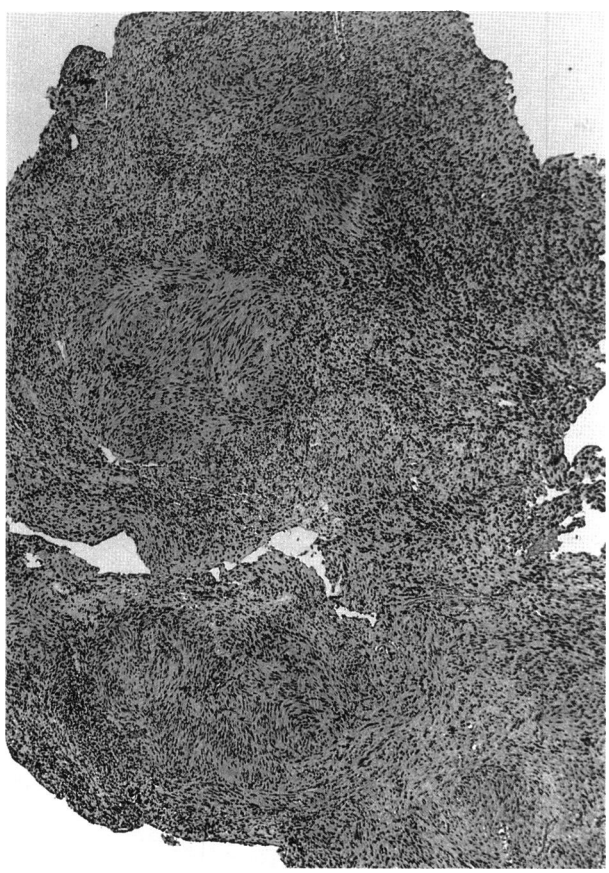

Figure 3. Histopathology of the resected lesion demonstrates a cellular pattern consistent with schwannoma.

computed tomography brain scan 2 months after surgery revealed minimal evidence of residual tumor in the cavernous sinus region (Fig. 4). Neuro-ophthalmological evaluation 3 months later demonstrated right third and fourth nerve pareses; however, the patient could fuse images well in primary gaze. Dilantin was discontinued 1½ years later after obtaining an electroencephalogram that did not reveal evidence of an epileptogenic focus.

The patient's progress was followed using serial magnetic resonance imaging scans, and no changes were noted until 34 months after surgery when enlargement of the right cavernous sinus lesion was noted. At the age of 16 the patient began to have increasing difficulty with diplopia. This progressed to the point where he was unable to fuse images in primary position, despite what appeared to be full extraocular movements. Repeat magnetic resonance imaging scans during the patient's 17th year of age revealed clear evidence of enhancing tumor extending from the superior cavernous sinus into the right sylvian fissure (Fig. 5). Neurological examination at that time was significant for right third and fourth nerve pareses. No motor deficits were noted. The patient was taken back to the operating room on June 20, 1989, 3 years and 10 months after his first surgery for resection of the enlarging residual tumor through the previous right frontotemporal craniotomy. Tumor seemed to extend from the region of an eroded anterior clinoid process along the dorsal and lateral aspects of the cavernous sinus. The third nerve was found to take a very medial course to its entry into the cavernous sinus which was encased by tumor and scar. Laser-assisted

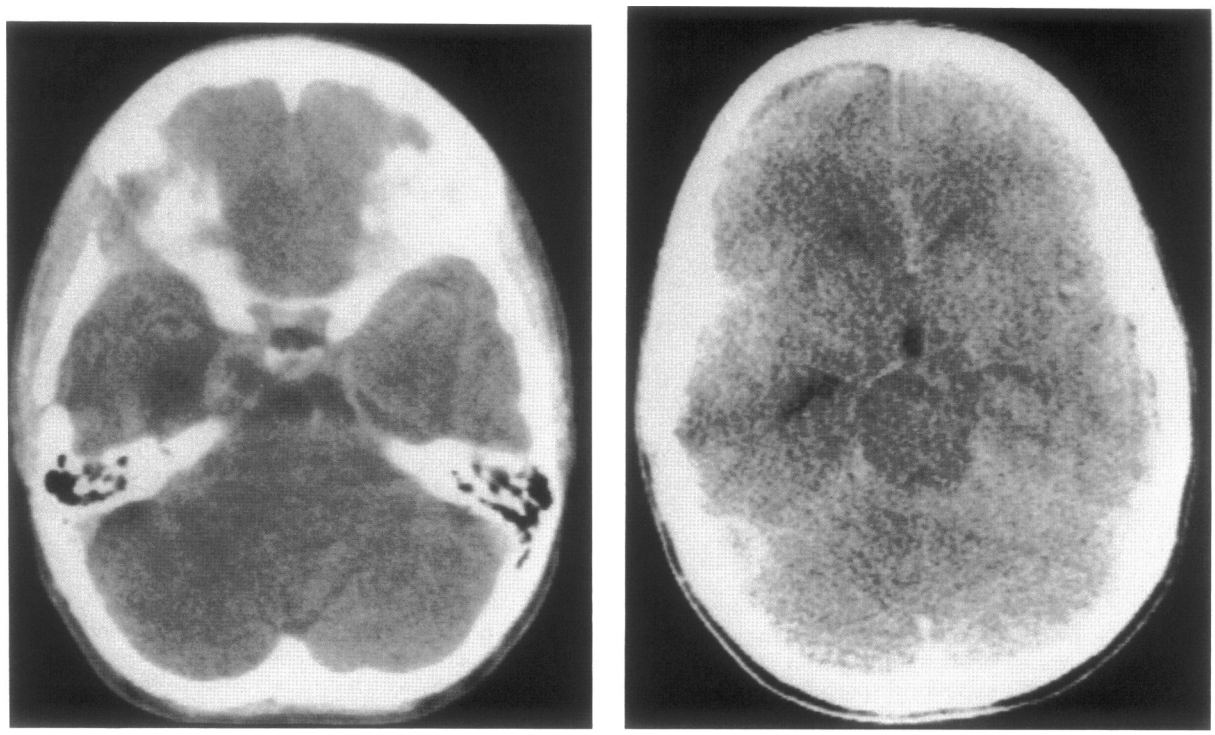

Figure 4. Computed tomography scan 2 months after the first operation (in 1985). A: Encephalomalacia in the region of right medial temporal lobe with no gross evidence of residual tumor. B: Computed tomography cut at a higher level demonstrating normal position of midbrain with no evidence of tumor in the posterior fossa.

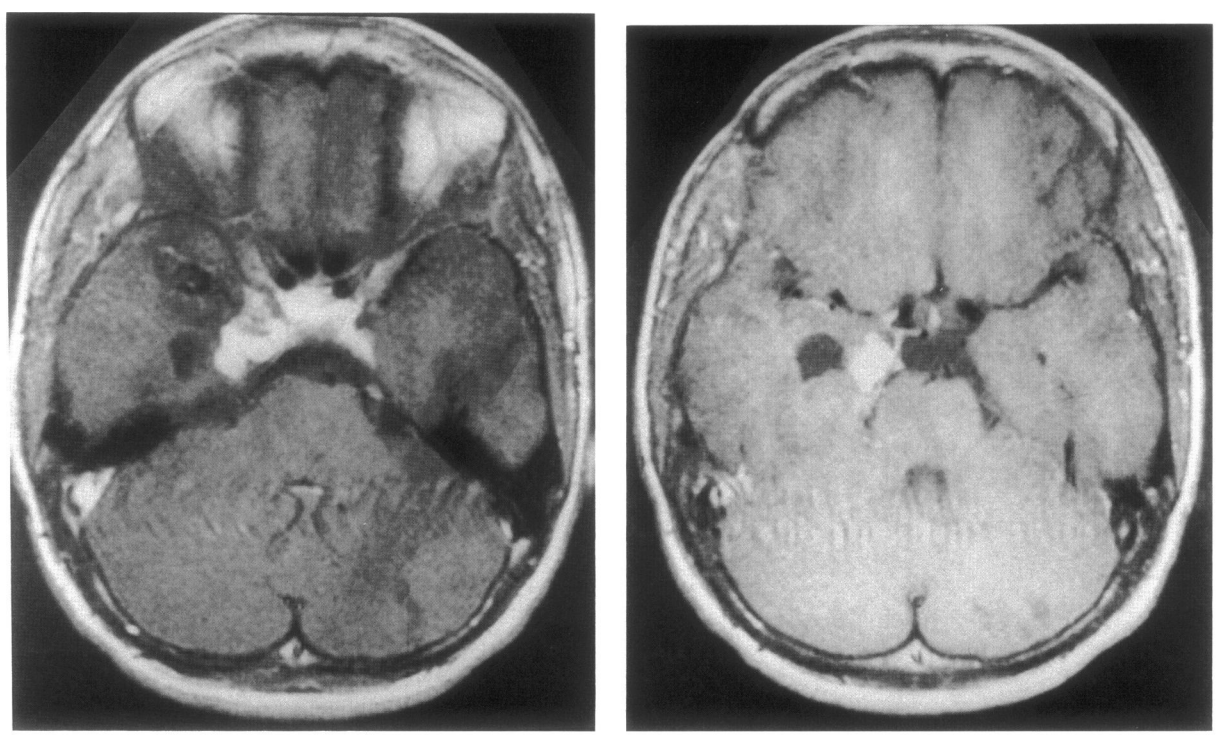

Figure 5. Follow-up magnetic resonance imaging with gadolinium in 1989. A: Enhancing tumor extending from the lateral wall of cavernous sinus. B: Upward extension of tumor mass in the area of previously observed encephalomalcia.

gross total removal of the tumor was then performed. Dissection along the free edge of the tentorium revealed that the fourth and fifth nerves were free of tumor. Postoperatively the patient had diplopia consequent to third and fourth nerve pareses. He did well otherwise. During the next 3 months the patient had further resolution of his fourth and third nerve pareses, and his diplopia resolved. The patient has continued to do well and is now 22 years old. The patient was reevaluated in November 1992 for symptoms of intermittent headaches, dizziness, and decreased visual acuity. His neurological examination showed mild paresis of muscles supplied by the third nerve without diplopia or ptosis. The reactive right pupil is larger than the left. The most recent magnetic resonance image of the brain in November 1992 has not shown any sign of tumor recurrence (Fig. 6). An angiogram was also obtained after the above magnetic resonance image raised a question of pseudoaneurysm of the right internal carotid artery. The latter study demonstrated slight dilatation of the posterolateral aspect of the supraclinoid internal carotid artery.

DISCUSSION

Isolated schwannomas account for about 8% of all primary intracranial neoplasms.[1] This tumor has a tendency to selectively involve sensory nerves and to arise on the distal segment of the nerve root after it has penetrated the pia mater and become insulated by Schwann cell–produced myelin. With the exception of the first and second nerves, cranial nerves are myelinated by Schwann cells distal to the Obersteiner-Redlich zone that is found within a few millimeters of the nerve root exit from the spinal cord or brainstem.[2] In general, schwannomas can arise at any age, but they are extremely rare in childhood.[3] There appears to be no sex predilection.[4] Acoustic schwannomas represent the overwhelming majority of these lesions that involve the intradural cranial nerves. Next in frequency are those arising from the trigeminal root or Gasserian ganglion, which constitute 0.2% of all intracranial tumors.[5] Rarely, these lesions arise in association with the glossopharyngeal (28 reported cases),[6] hypoglossal (16 reported cases),[7] facial,[8] or vagus nerves. Schwannomas originating from cranial nerves innervating the extraocular muscles are extremely rare in the absence of neurofibromatosis type I. Eighteen percent of solitary intracranial schwannomas occur in the presence of neurofibromatosis.[4] A review of the world's literature discloses 9 reports of schwannomas involving the abducent nerve, 11 of schwannomas of trochlear nerve origin, and 26 cases arising from the oculomotor nerve.

Schwannomas should not be confused with neurofibromas, which tend to have different histologic features and behavior. Macroscopically, schwannomas are soft, discrete, sometimes yellow, and may contain cystic areas. Microscopically, lesions are variably composed of bundles of spindle cells, called Antoni type A cells, and looser

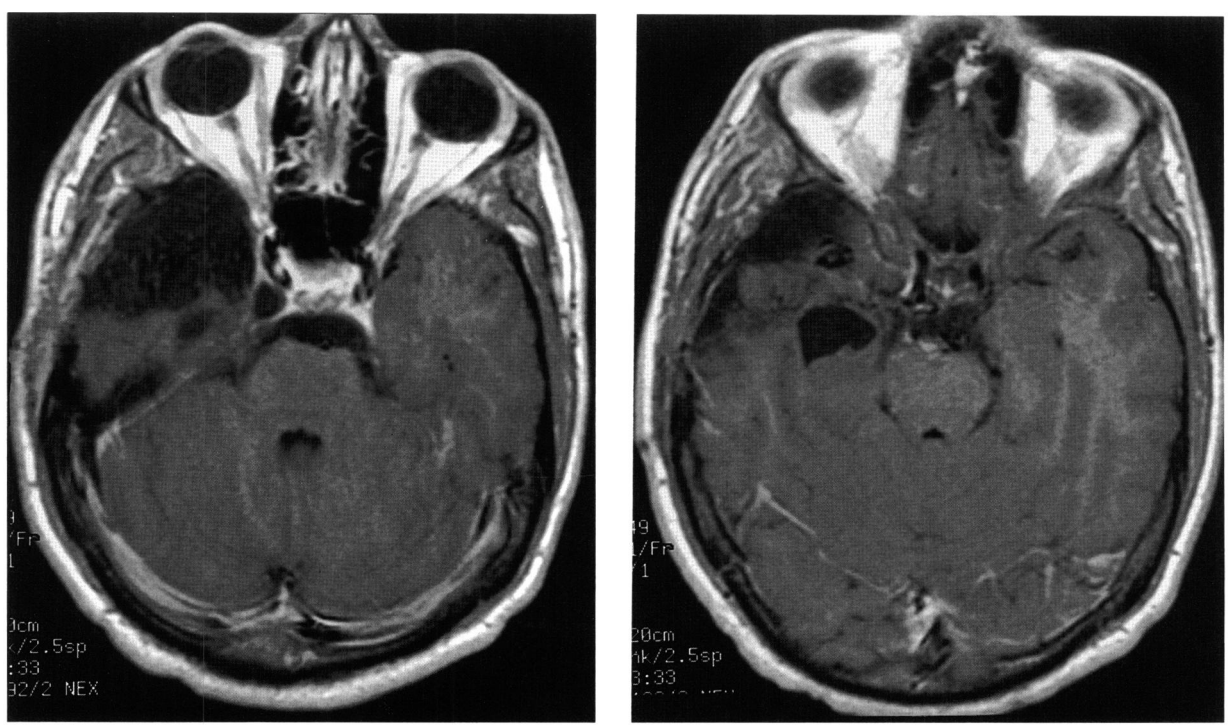

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Figure 6. Magnetic resonance imaging scan with gadolinium in November 1992 (3½ years after the resection). A: Area of hypointensity without enhancement in the right parasellar region. B: Area of hypointensity without enhancement in the right medial temporal lobe consistent with encephalomalcia and no evidence of residual tumor.

elements producing a spongy appearance, called Antoni B cells. Both these cell types grow Schwann cells on tissue cultures.[9] Schwannomas are most frequently benign slow-growing solitary lesions that are encapsulated, conforming to the shape of the space in which they grow, and compress rather than invade the normal tissue. Unlike neurofibromas, they tend to arise from a discrete location of the nerve sheath and displace rather than interdigitate with the associated nerve fibers. Schwannomas can often, therefore, be dissected from the nerve without compromising its function. When these lesions are incompletely removed, there is almost no danger of their becoming malignant and metastasizing.[10] In contrast, neurofibromas tend to be multiple; grow within the endoneural substance, rendering them difficult to dissect; and may undergo malignant degeneration.[11]

The fact that the extraocular and trigeminal nerves are closely related in the middle cranial fossa often makes it difficult to determine the site of tumor origin. It is also possible that schwannomas can arise from minute nerves innervating the dura mater. Review of the literature on schwannomas arising from nerves supplying the extraocu-

Table 1. Summary and Sequences of Cranial Nerve Involvement of Reported Cases of Schwannomas of Extraocular Nerves

Nerve of Origin	*Author*	*Age*	*Sex*	*Duration*	*Symptoms*
III	Kovacs[13] (1927)	55	M	Incidental	
	Nagamune et al[14] (1974)	46	F	?	III
	Shuangshoti[15] (1975)	64	F	Autopsy	II–"III"
	Kan et al[16] (1976)	36	M	?	"III", V, exophthalmos
	Huber[17] (1978)	40	F	10 y	II–"III"
		54	F	13 y	II–"III" and IV
		52	M	6 mo	"III", IV, VI, V
	Wakabayashi et al[18] (1978)	70	M	Incidental	
		40	F	?	II, "III", exophthalmos
	Schubiger et al[19] (1980)	19	F	2 y	"III"
	Broggi and Franzini[20] (1981)	55	M	5 y	III
	Sako et al[21] (1981)	29	F	8 mo	II, III, exophthalmos
	Hiscott and Symon[22] (1982)	58	F	6 mo	Hemiparesis, "III"
	Kansu[23] (1982)	15	M	11 y	III
	Leunda et al[24] (case 2, 1982)	11	M	3 mo	III, hemiparesis
	Okamoto[25] (1985)	52	F	18 mo	III, exophthalmos
	Ishige et al[26] (1985)	64	F	1 mo	"III", retro-orbital pain
	Satoh et al[27] (1985)	53	F	?	III
	Nogami et al[28] (1986)	40	F	6 y	III
	Bataille et al[29] (1987)	49	F	5 y	II, "III", IV, VI, exophthalmos
	Katsumata et al[30] (1990)	47	M	4 mo	"III"
	Lunardi et al[31] (1990)	60	F	1 y	III, hemiparesis
	Takano et al[32] (1990)	65	F	2 mo	III
	Mehta et al[33] (1990)	19	F	9 mo	III, cerebellar signs
	Kurokawa et al[34] (1992)	55	F	2 wk	III
	Barat et al[35] (1992)	27	F	16 y	"III", IV, VI, exophthalmos
IV	King[36] (1976)	55	F	3 mo	V contralateral
	Boggan et al[37] (1979)	32	F	2 yr	V–"IV"
	Ho[38] (1981)	58	F	Incidental/autopsy	
	Samii[39] (1981)	?		?	IV
	Leunda et al[24] (1982)	54	M	7 mo	"IV"–III
		16	F	5 mo	"IV"
	Garen et al[40] (1987)	18	F	6 mo	III
	Yamamoto et al[41] (1987)	37	F	2 wk	IV
	Tokuriki et al[42] (1988)	43	M	13 mo	V (taste), hemiparesis and sensory disturbances
	Maurice-Williams[43] (1989)	56	M	5 mo	IV, spastic hemiparesis
	Celli et al[44] (1992)	51	M	12 mo	IV, ataxia, hemiparesis
VI	Chen[45] (1981)	46	F	2.5 mo	Raised ICP–"VI"
	Leunda et al[24] (1982)	10	M	2 mo	"VI"
	Hansman et al[46] (1986)	58	M	Acute	"VI"
	Ginsberg et al[47] (1988)	47	F	2 y	"VI", VII, raised ICP
	Nehls et al[48] (1985)	38	M	3 mo	VI
	Tung et al[49] (1991)	35	M	1 y	"VI"
		45	F	3 y	"VI"
	Lanotte et al[50] (1992)	62	M	?	III, VI
	Barat et al[35] (1992)	49	F	14 mo	III, VI

ICP = intracranial pressure.

lar muscles (Table 1) suggests that the initial symptom due to cranial nerve involvement is an important clue as to the origin of the tumor. In 46 reported cases of schwannomas of the extraocular nerves, the most common symptom was headache, followed by diplopia. Less than one third of the cases had involvement of other cranial nerves on presentation. An associated hemiparesis was noted in 5, exophthalmos in another 6, and cerebellar signs in 2 cases. The third nerve is rarely affected by the tumors compressing or arising within the cavernous sinus. Approximately 90% of patients harboring trigeminal schwannomas have clinical evidence of sensory or motor involvement of the trigeminal nerve at the time of diagnosis.[12] In contrast to the authors's case, schwannomas of the trigeminal nerve classically displace the superior cerebellar and anterior inferior cerebellar arteries in opposite directions. The separation of the posterior cerebral and superior cerebellar arteries by the tumor strongly corresponds to the location of the third cranial nerve.

The case reported herein is relatively unusual because of the patient's young age and ipsilateral hemiparesis at the time of presentation. His postoperative neurological deficits of third and fourth cranial nerve pareses were likely due to compression by tumor and injury to these nerves during the operative dissection. Although the exact origin of this tumor, as in other case reports reviewed, could not be ascertained precisely at the time of first operation, pre- and postoperative findings strongly suggest a third nerve origin. Because all cases reporting surgically resected schwannomas involving the extraocular nerves have had reasonable results, the authors believe an aggressive surgical approach to these lesions is warranted.

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