Sixth nerve schwannomas

Report of two cases

HOWARD TUNG, M.D., THOMAS CHEN, M.D., AND MARTIN H. WEISS, M.D.

Department of Neurological Surgery, University of Southern California School of Medicine, Los Angeles, California

Two cases of sixth cranial nerve schwannoma are presented with a review of four other cases from the literature. The clinical spectrum, neuroradiological findings, and surgical outcome of the six cases are discussed. There are two distinct clinical presentations for sixth cranial nerve schwannomas. Type I sixth nerve schwannomas present with sixth nerve palsy and diplopia and arise from the cavernous sinus. In contrast, type II sixth nerve schwannomas have a more severe presentation with obstructive hydrocephalus, raised intracranial pressure, sixth nerve palsy, and diplopia. This type arises along the course of the sixth cranial nerve in the prepontine area. Cavernous sinus involvement in either type may preclude total surgical excision and indicate an increased possibility for recurrence.

KEY WORDS • schwannoma • sixth cranial nerve • cavernous sinus • prepontine area

Schwannomas (neurinomas or neurilemomas) constitute approximately 8% of all intracranial neoplasms and most commonly involve the fifth or eighth cranial nerves.11 Schwannomas of the sixth cranial nerve are exceptionally rare; only four cases have been previously reported.14,5,7 We report two patients with sixth nerve schwannoma in the absence of neurofibromatosis and review the literature of this rare clinical and anatomical entity.

Case Reports

Case 1

This 35-year-old man with a 1-year history of diplopia on left lateral gaze was examined in February, 1990. He denied experiencing headaches or other neurological symptoms.

Examination. On neurological examination, only a left sixth nerve palsy could be demonstrated. Magnetic resonance (MR) imaging was obtained with multiplanar views of T1-weighted, T2-weighted, and gadolinium-enhanced T1-weighted images. A 2.0-cm cystic lesion was identified mainly involving the left cavernous sinus and parasellar region. A solid component also extended to the interpeduncular region. The lesion appeared as a low-intensity area on T1-weighted images and as hyperintense on T2-weighted images. The mass was noted to be displacing the carotid artery medially. Gadolinium-enhanced images revealed the mass to be ring-enhancing with a low-intensity center, confirming the cystic nature of the lesion (Fig. 1). A preoperative diagnosis of fifth nerve schwannoma was made.

Operation. The patient underwent a frontotemporal craniotomy with microexcision of the tumor. The lesion was noted to be cystic and arising from the sixth cranial nerve which was thickened. The third, fourth, and fifth cranial nerves were identified during surgery and appeared to be normal. Neuropathological examination showed the tumor to be a typical schwannoma. Postoperatively, the patient did extremely well but experienced third, fourth, and sixth nerve palsies of which the third and fourth nerve palsies quickly resolved. Follow-up MR imaging confirmed total excision of the tumor from the parasellar region and cavernous sinus.

Case 2

This 45-year-old woman with a 3-year history of diplopia was referred to our service in May, 1990, because of a new onset of mild headache.

Examination. On neurological examination, she manifested a left sixth nerve palsy. An MR image revealed a 3.2-cm low-intensity lesion, with multiple
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![Image 1: Case 1. Left: Sagittal T1-weighted magnetic resonance (MR) image showing a low-intensity parasellar mass with extension to the preoptic area along the course of the sixth cranial nerve. Right: Gadolinium-enhanced coronal MR image showing the parasellar mass to be ring-enhancing and demonstrating the cystic nature of the lesion.](image1)

![Image 2: Case 2. Left: Axial T2-weighted magnetic resonance (MR) image revealing a parasellar mass with multiple cystic areas. Right: Gadolinium-enhanced coronal MR image showing intense enhancement, highlighting the multiple cystic areas within the lesion.](image2)

small cystic areas involving the left parasellar region visualized on the weighted images. The mass was hyperintense on the T2-weighted images. Gadolinium-enhanced MR images showed uniform enhancement of the lesion and accentuated the multiple cystic areas within it (Fig. 2). The tumor was noted to be displacing and encasing the carotid artery in the cavernous sinus. A preoperative diagnosis of fifth nerve schwannoma was made.

Operation. The patient underwent a frontotemporal craniotomy with microsurgical resection of the tumor. The tumor was excised in all areas except the cavernous sinus where it densely adhered to and circumferentially surrounded the carotid artery. At surgery, multiple small cysts were noted within the lesion and the tumor was identified to be arising from the sixth cranial nerve. Neuropathological examination revealed a schwannoma. Postoperatively, the patient did very well. She demonstrated palsies of the third, fourth, and sixth cranial nerves and of the first division of the trigeminal nerve, all of which resolved quickly, except for the sixth nerve palsy which remained at her follow-up examination. Follow-up MR imaging 6 months after surgery showed residual tumor within the cavernous sinus.

Discussion

With the exception of the first and second cranial nerves, all the cranial nerves have myelinated sheaths composed of Schwann cells and are potential sites for intracranial schwannomas.\(^1\)\(^2\)\(^1\) Schwannomas account for approximately 8% of all primary intracranial tumors\(^3\)\(^4\)\(^5\) and, furthermore, approximately 18% of solitary intracranial schwannomas occur in the presence of neurofibromatosis.\(^6\) Schwannomas have a distinct propensity to affect sensory nerves more than pure motor nerves. The vestibular division of the eighth cranial nerve is the most commonly affected, with the trigeminal nerve root or ganglion next in frequency of occurrence.\(^7\) Schwannomas arising from other cranial nerves are very rare.

Anatomical Considerations

The sixth cranial nerve (abducens nerve) arises from the floor of the fourth ventricle close to the midline and beneath the facial colliculus. The fibers of the sixth nerve pass anteriorly and caudally through the pons to emerge at the pontomedullary junction. The sixth nerve then courses rostrally between the pons and the anterior inferior cerebellar artery along the clivus in the subarachnoid space, penetrating the dura below the posterior clinoid processes, to enter the cavernous sinus. In the cavernous sinus, the sixth nerve is situated in the lumen of the sinus away from the wall. The nerve then enters the orbit through the superior orbital fissure to innervate the lateral rectus muscle from its medial side.\(^8\)

Clinical Spectrum

Including our two cases, six cases of sixth cranial nerve schwannoma have been identified (Table 1).\(^9\)\(^1\)\(^2\)\(^4\)\(^5\)\(^7\) The age of the patients at diagnosis was variable and ranged from 10 to 58 years. There were three males and three females and an even distribution of right- and left-sided lesions. No association with neurofibromatosis was identified.

The clinical presentation included cranial sixth nerve palsy and diplopia in all cases; headache was present in four cases. Obstructive hydrocephalus with signs and symptoms of raised intracranial pressure were observed in three patients. The duration of symptoms was variable, with a mean period of approximately 13 months, but tended to be shorter in patients presenting with hydrocephalus.

In our two cases, the presenting symptoms were sixth nerve palsy with diplopia and without hydrocephalus. In both cases the neoplasm arose from the cavernous sinus. There appear to be two distinct clinical presentations of sixth cranial nerve schwannoma depending
upon the site at which the neoplasm arises and its predominant location along the course of the sixth cranial nerve. If the sixth cranial nerve schwannoma arises from the cavernous sinus (type I), the clinical symptoms include only sixth nerve palsy and diplopia with or without mild headache. In contrast, if the sixth cranial nerve schwannoma arises from and predominantly occupies the preptontine area before entering the cavernous sinus (type II), the presentation is more malignant. The tumor is more likely to grow and manifest symptoms of obstructive hydrocephalus prior to or in tandem with diplopia and sixth nerve palsy. In addition, other cranial nerve findings may also be observed (Table 1).

Neuroradiological Imaging

The majority of intracavernous lesions seen on radiological imaging are meningiomas and aneurysms.14 The MR imaging characteristics of intracranial schwannomas have been well described3,8,9 and a preoperative diagnosis of schwannoma was made in both of our cases based on this method. However, preoperative differentiation between sixth nerve and fifth nerve schwannomas can be extremely difficult. Fifth nerve schwannomas have been reported to present initially with sixth cranial nerve palsy,3,15 but these lesions originate from the nerve root in the cerebellopontine angle or from the trigeminal ganglion in Meckel's cave and not from the cavernous sinus, as in our two cases. Cavernous sinus masses usually have a broad attachment laterally to the cavernous sinus and often show a tapering block of normal brain inferiorly between the tumor and the base of the middle fossa (Figs. 1 right and 2 right).12 On the other hand, tumors originating from the fifth nerve ganglion in Meckel's cave would be expected to have a broader attachment to the base of the middle fossa. Finally, the tumor in Case 1 was seen to be growing along the course of the sixth cranial nerve (Fig. 1 left) and was noted to be distinct from the gasserian ganglion, which was normal in appearance on MR imaging.

Surgical Outcome

Surgical resection has been attempted in all six reported cases, including our two. The surgical corridor chosen will depend on the exact anatomical location of the neoplasm in each case. With type I sixth nerve schwannomas involving predominantly the cavernous sinus and parassellar region, the frontotemporal transcavernous approach may be the most advantageous. For type II sixth nerve schwannomas predominantly involving the preptontine area, the lateral transtentorial or subtentorial approach may be more appropriate. Despite the more malignant clinical presentation and generally larger size of type II sixth cranial nerve schwannomas, these lesions may afford a better opportunity for total surgical excision, especially if the cavernous sinus is spared. The two cases from the literature in which total excision was achieved were of this variety.2,3

Although total microneurosurgical excision with preservation of neurological function is the goal of surgery, involvement of the cavernous sinus in either type of sixth cranial nerve schwannoma may preclude total surgical excision, as separation of the neoplasm from the vital structures of the cavernous sinus may be impossible without unacceptable neurological morbidity. This was apparent in one of our two cases and in some previous cases reported in the literature.4,5 Furthermore, since the potential for recurrence of sixth
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cranial nerve schwannomas involving the cavernous sinus is theoretically higher, these patients may be candidates for postoperative conventional or stereotactically directed radiation therapy.

References

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Address reprint requests to: Martin H. Weiss, M.D., Department of Neurological Surgery, LAC-USC Medical Center, 1200 North State Street, Room 5046, Los Angeles, California 90033.