Neurilemmoma of the fourth cranial nerve

Case report

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A tumor of the trochlear nerve sheath with an unusual but diagnostic presentation is described. The rarity of reported cases may reflect failure to differentiate tumors originating from the trochlear and trigeminal nerves.

KEY WORDS • trochlear nerve • neurilemmoma

Nerve-sheath tumors comprise less than 8% of all intracranial neoplasms, and usually involve sensory nerves. To our knowledge, there are only two reported cases, unassociated with von Recklinghausen’s disease, of neurilemmomas of the cranial nerves innervating the extraocular muscles. This report presents the case of a patient with a trochlear nerve neurilemmoma, and a review of the literature relative to this rare tumor.

Case Report

History. In September, 1975, 2 months after a terminated pregnancy and a tubal ligation, this 32-year-old right-handed woman had the onset of constant right-sided retro-orbital headaches, which were attributed to emotional stress. After approximately 6 weeks, the patient experienced the onset of double vision. Examination by an ophthalmologist revealed a right superior oblique palsy, but her neurological examination was otherwise normal. A tentative diagnosis of Tolosa-Hunt syndrome was made, and the patient was started on corticosteroids, which relieved her double vision, but did not alleviate her headaches. When the use of corticosteroids was discontinued, her diplopia returned. Extensive work-up, including computerized tomography (CT) brain scans, cerebral angiography, skull films, and lumbar puncture, revealed no abnormality. A diagnosis of migraine headaches was made. The patient was treated with ergonovine with relief of her headaches, but her diplopia persisted and she required increasing dosages for maintenance of pain relief.

In February, 1977, the patient experienced a transient episode of right facial weakness, numbness, and tingling that resolved spontaneously. Approximately 2 months before admission, she noted increasing left-sided weakness and a lack of coordination with a tendency to fall to the left side. In addition, she experienced numbness and tingling on the left side of her tongue and face, and in her left hand. The patient also experienced frequent episodes of nausea, occasional drooling from the left side of the mouth, and amenorrhea.

Examination. In September, 1977, the patient was admitted to the Neurological Surgery Service, University of California, San Francisco. She appeared to be in good health, and her higher cortical functions were intact. Disc margins, visual fields, and acuity were normal. Anisocoria was noted, with the right pupil larger than the left, which the patient recalled had always been the case. Both eyes reacted briskly to light. A right fourth-nerve palsy was present, and the gag reflex appeared slightly depressed bilaterally. The other cranial nerves were normal. Left-sided hemiparesis, hyperreflexia, and Babinski and Hoffmann signs were noted. Her gait was broad-based. She tended to fall to the left with tandem walking and demonstrated left appendicular dysmetria. There were no cutaneous manifestations of von Recklinghausen’s disease.

Anteroposterior and lateral polytomograms of the skull base revealed no evidence of bone erosion or
hyperostosis, and the petrous bones were normal. An isotope brain scan (\textsuperscript{99m}Tc) revealed a deep right-sided paramedial lesion. Coronal and axial CT scans revealed a solitary enhancing mass in the region of the right tentorial incisura, with supra- and infratentorial extension (Fig. 1). The lateral ventricles were slightly enlarged. Right external and internal carotid (Fig. 2) and vertebral angiography (Fig. 3) revealed the presence of a right incisural vascular mass. The preoperative diagnosis was right tentorial meningioma.

**Operation.** A right frontotemporoparietal osteoplastic craniotomy was performed (C.B.W.). After retraction of the temporal lobe, the tumor was seen to be elevating the tentorium. Incision into the dura revealed a soft yellow tumor with an overall diameter of 4 cm. Following resection of the superior aspect of the tumor, the capsule was teased from the compressed right cerebral peduncle and sharply dissected from the posterior cerebral artery. The third nerve was minimally stretched over the medial and inferior aspect of the tumor. The fourth nerve entered directly into the tumor posteroinferiorly and exited from the tumor just in front of its entrance into the cavernous sinus. The Gasserian ganglion and fifth nerve were displaced laterally, but were clearly unattached to the tumor. The tumor was totally excised.

**Postoperative Course.** At follow-up examination 8 months postoperatively, the patient has an absent right corneal reflex. She has required ophthalmological correction of her diplopia from persistent trochlear nerve paralysis. Otherwise, she is neurologically normal and fully active.

**Pathological Examination.** Examination of tumor specimens revealed characteristic spindle-shaped cells in areas of typical Antoni A and Antoni B patterns. Numerous Verocay bodies were present. Large vascular structures with irregular hyalinization of their walls were noted. There were numerous cystic spaces and sinusoids containing pale eosinophilic material and foci of fresh hemorrhage. There were also multiple foci of lymphocytic infiltrate. The tissue was characteristic of a neurilemmoma.

**Discussion**

An acquired isolated fourth-nerve palsy is most frequently associated with trauma. In approximately 8% of reported cases, the palsy is the result of secondary involvement by tumor; none has been primary to the trochlear nerve. Intracranial neurilemmomas most frequently arise from the vestibular nerve and less frequently from the trigeminal nerve or ganglion; they rarely involve a cranial motor nerve. These tumors occur twice as frequently in females as in males, and usually present clinically during the third and fourth decades of life. In the only reported case of a trochlear nerve neurilemmoma, the tumor's origin was less precisely defined than in the present case.

Because of the close anatomical relationship between the trigeminal and trochlear nerves, we reviewed the literature regarding trigeminal root and ganglion neurilemmomas. In several cases, diplopia was an early manifestation of the tumor, but the operative and postoperative findings did not support a nontrigeminal origin. We suggest, however, that some of these tumors, particularly those in which trigeminal involvement was inconspicuous on
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**References**


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