Trochlear nerve sheath tumor

Case report

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The author describes the presenting symptoms and treatment of a patient with a trochlear nerve sheath tumor.

KEY WORDS • trochlear nerve • nerve sheath tumor

To my knowledge, a trochlear nerve sheath tumor has not previously been reported. The following is a case of such a tumor, notable also because of the presenting symptoms.

Case Report

A 55-year-old woman was admitted complaining of right facial numbness of 3 months' duration. She had apparently been well until approximately 1 year prior to admission, when she suddenly became unsteady while walking, like "being on a ship at sea." She had to walk with her feet wide apart, and frequently bumped into things. This problem persisted for many months and then became intermittent. Three months prior to admission she noted the onset of a "pins and needles" sensation over her right cheek which gradually spread to involve the skin over the parotid gland and the supraorbital scalp. She then discovered that she could not feel touch over the right side of her face. The area of hypesthesia gradually spread to involve her right arm and leg. During the month before admission she was bothered by intermittent occipital headaches, and 2 weeks prior to admission she experienced an acute attack of vertigo which forced her to remain in bed for several hours.

Examination. The patient was a pleasant and cooperative middle-aged woman of slight build. She was well-oriented, subtracted 7's from 100 without error, and recalled recent and past events without difficulty. Her visual acuity was 20/30 bilaterally, her discs were flat, and visual fields were full to confrontation. The extraocular movements were full and she denied diplopia. She had a depressed right corneal reflex and subjectively decreased sensation of touch over the area supplied by the first and second divisions of the fifth cranial nerve on the right. A mild right central facial palsy was present. She heard finger rub bilaterally. She swallowed on command and protruded her tongue in the midline. During motor testing a mild drift of the right arm was noted, but weakness appeared to be minimal in individual muscle tests. Deep tendon reflexes were slightly more active on the right and the right plantar reflex was extensor. Gait was normal. Finger-to-nose testing showed dysmetria on the right.
but heel-to-shin testing was normal bilaterally. Rapid alternating movements of the right arm were slightly irregular. Somesthetic testing of the extremities was normal.

Lumbar puncture yielded an opening pressure of 180 mm H₂O, and the cerebrospinal fluid protein was 93 mg. A brain scan showed a spherical focus of ¹⁹⁵Tc accumulation in the midline suboccipital region. Bilateral carotid and vertebral angiograms indicated displacement of the left superior cerebellar artery above the posterior cerebral artery. A pneumoencephalogram revealed a 3 × 4-cm extraaxial mass at the incisura on the left.

Operation. On the fifteenth day after admission a left temporoparietal osteoplastic craniotomy was performed. Elevation of the left temporal lobe disclosed the tumor elevating the tentorium. It was located just lateral to the incisura and measured approximately 3 cm in diameter. The tentorium was incised over the mass, and the tan-colored soft encapsulated tumor was removed piecemeal. The tumor was not adherent to the tentorium. The posteroinferior aspect of the tumor was dissected from the trochlear nerve, which was its only nerve attachment; it did not involve the tentorium other than by displacement. The tumor was removed totally except for a very small portion which was firmly fixed to a large artery entering the brain stem in this area.

Pathological Examination. The tumor specimens were examined by a neuropathologist, who noted two distinct histopathological patterns (Fig. 1). Very compact cellular areas alternated with less prominent regions showing a loose tissue pattern. Under higher power the spindle-shaped cells that made up the majority of the tumor could be seen arranged in bundles, which streamed in many directions. Conspicuous areas that suggested palisading of the oval or fusiform nuclei were apparent. Mitotic activity was absent and nuclear pleomorphism was minimal. Numerous very thick-walled, hyaline vessels were seen throughout the tumor tissue. The distinctive two-pattern morphology, palisading in the more compact areas, and the typical rich, delicate reticulin pattern observed after silver staining are unequivocal characteristics of a nerve-sheath tumor under light microscopy.

Postoperative Course. Postoperatively the patient’s preoperative signs and symptoms abated. In the immediate postoperative period she was seen in consultation by a neuroophthalmologist who noted a mild left
superior oblique palsy. While in the hospital the patient was started on radiotherapy which was then continued on an outpatient basis. She received 5500 rads over a 6-week period.

Upon re-examination 18 months later she reported that none of her previous problems had recurred, although she had persistent diplopia upon looking down and to the right. She demonstrated a left superior oblique palsy and minimal proximal weakness of her right leg. Babinski reflex could still be elicited on the right.

Discussion

Primary intracranial nerve sheath tumors are usually discovered in the middle decades of life and are far more common in females than males. In cases other than von Recklinghausen’s disease, such tumors have, to our knowledge, always involved the sensory nerves. Most of these have come from the vestibular nerve (not the acoustic nerve), but others have been reported originating from the trigeminal nerve, most frequently from the Gasserian ganglia. Russell and Rubinstein have reported an incidental autopsy finding of a schwannoma on the ninth root, and Strum, et al., have reported one from the olfactory nerve.

To the author’s knowledge, the case presented here constitutes the first report of a solitary nerve-sheath tumor originating from the trochlear nerve and the first report of evolution of such a tumor from a purely motor cranial nerve. It is difficult to explain the constellation of right-sided symptoms and signs in this case on the basis of mass located at the left incisura, particularly because of the depressed right corneal reflex and the hypesthesia over the first two divisions of the trigeminal nerve. Possibly the second-order neurons of the right trigeminal nucleus were interrupted as they traversed the left side of the mesencephalon. The right-sided motor signs would of course be anticipated from compression of the peduncle at this point.

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References

3. Hoyt WR: Personal communication, September, 1973

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