Trochlear Nerve Neurinoma

—Case Report—

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Abstract

A 43-year-old male presented with a low-density mass on computed tomographic scans; the mass did not exhibit contrast enhancement. The tumor was subtotally removed, and the intraoperative diagnosis was trochlear nerve neurinoma. Six other cases have been reported. The characteristic symptoms and signs of this tumor appear to be unusual facial sensations and signs of brain stem compression rather than trochlear nerve palsy.

Key words: neurinoma, trochlear nerve, brain tumor symptomatology

Introduction

Intracranial neurinomas are common brain tumors, surpassed in frequency only by gliomas, meningiomas, and metastatic tumors. Most neurinomas originate from the eighth cranial nerve, although neurinomas arising from the fifth, ninth, or twelfth cranial nerves are sometimes seen. However, neurinomas originating from pure motor nerves, such as the third, fourth, or eleventh cranial nerves, are extremely rare. We were able to find only nine,⁶⁾ six,⁸⁾ and five^{2,7)} cases, respectively, in the literature. We report a case of trochlear nerve neurinoma in which the connection between the trochlear nerve and the tumor was confirmed at operation.

Case Report

A 43-year-old, right-handed male was admitted to our department with a chief complaint of gait and left-side sensory disturbances. Thirteen months before admission, he had noticed numbness in his left hand and foot. A few months later he became unsteady while walking and insensible to touch on the left side of his body. He consulted a physician and received some drugs, but his condition gradually de-

teriorated. On February 13, 1985, 2 weeks prior to admission, he had an acute episode of left hemiplegia, which improved within a few days. He had no headaches or diplopia.

The physical examination showed the patient to be in good general health. The neurological examination was normal except for disturbance of all senses and mild motor weakness on the left side. He had no eye movement disorders. His symptoms advanced, and he began to experience numbness and tingling in the left face and disturbance of taste. There were no cutaneous manifestations of von Recklinghausen's disease.

Plain skull x-rays revealed no abnormalities, and Towne's view disclosed no erosion of the petrous bone. Computed tomography (CT) demonstrated a low-density mass with clear margins in the region of the right tentorial incisura, with the main portion located in the infratentorial region (Fig. 1A). This mass was not enhanced by contrast medium (Fig. 1B). Coronal CT also revealed a low-density area, which occupied almost one half of the midbrain (Fig. 1C). The initial diagnosis was epidermoid tumor. Metrizamide CT scan showed that the ambient cisterns were symmetrical and that over one half of the midbrain region was filled with a low-density mass sug-

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Fig. 1 A, B: CT scans showing a clearly demarcated, low-density mass in the right midbrain (A), which is not enhanced after the administration of contrast medium (B). C: Coronal CT scan also showing a low-density area occupying almost one half of the midbrain. D: Metrizamide CT scan showing the symmetrical ambient cistern, which suggests an intra-axial mass lesion.

gestive of an intra-axial lesion (Fig. 1D). Magnetic resonance imaging (MRI) revealed a clearly demarcated extra-axial cystic mass adherent to the midbrain, which was displaced dorsally and to the right (Fig. 2). Vertebral angiography showed downward deviation of the superior cerebellar artery. The posterior cerebral artery was in the normal position but appeared partially obstructed (Fig. 3 *upper*). Vascular staining was negative (Fig. 3 *lower*).

On March 20, 1985, through a right temporal craniotomy, the tumor was approached *via* the subtemporal transtentorial route. The tentorium was elevated by a dark yellow, encapsulated tumor. Puncture of the tumor yielded about 6 ml of cystic fluid. The tumor was well demarcated from the brain stem. Near the cranial pole of the tumor capsule, the posterior cerebral artery and oculomotor nerve were identified. The trochlear nerve entered directly into the tumor posteromedially. As the tumor adhered tightly to the dura covering the petrous tip and caver-

nous sinus, the incisura tentorii and the exit site of the trochlear nerve could not be confirmed. The superior cerebellar artery adhered to the inferior surface of the tumor capsule. The tumor was subtotally removed; the portions adherent to the superior cerebellar artery and lateral aspect of the pons could not be excised.

Postoperatively, he developed diplopia due to trochlear nerve palsy. However, the left motor deficit and sensory and taste disturbances disappeared completely within 1 month. A postoperative CT scan revealed a normally positioned brain stem.

Microscopically, the tumor cells contained compact, interlacing bundles of elongated, bipolar, spindle-shaped nuclei, some of which exhibited palisading typical of Antoni type A neurinoma. There was a small area in which the cells were arranged in an indefinite pattern (Antoni type B). Hyalinized vascular structures were also noted. The histological diagnosis was neurinoma (Fig. 4).

Discussion

Neurinomas of the cranial pure motor nerves are extremely rare. The first case of trochlear nerve neurinoma was reported by King⁵⁾ in 1976, and we found

Fig. 2 MRI [superconducting system (0.5 tesla) of Shimadzu]. A: IR image (TR = 1500 msec, TI = 500 msec, TE = 25 msec) showing a round area of low signal intensity in the right ponto-mesencephalic junction. This abnormal low-intensity area has a clear margin and appears to compress the right side of the pons and the cerebral peduncle. The brain stem is displaced leftward. The ambient cistern is open on the right side. B: In the SE image (TR = 2000 msec, TE = 80 msec), the lesion appears as an area of markedly high signal intensity, which suggests that the T_2 of the lesion is very long.

Fig. 3 Right vertebral angiography. *upper*: Anteroposterior views of the arterial phase show inferior displacement of the right superior cerebellar artery. The right posterior cerebral artery filled poorly but was almost normally positioned. *lower*: Lateral views of both the arterial (*left*) and venous (*right*) phases show no tumor staining.

only five subsequent cases in the literature^{1,3,6,8)} (Table 1). One of the tumors (Case 3) was found incidentally at autopsy. The patients' ages ranged from 16 to 58 years; two of the patients were males and

five were females.

Headache and diplopia were the most common symptoms, although such information was not available in Case 3 and two cases were not described in detail. Of the remaining four patients, three showed no trochlear nerve palsy. Cases 1 and 2 were similar to our case in that there were symptoms of compression of the cerebral peduncle or mesencephalon, *i.e.*, contralateral sensory disturbance and hemiparesis. Perhaps most specific was abnormal sensations in the contralateral face, described as "pins and needles" in Case 1 and as "tingling" in Case 2. Our patient also experienced odd sensory phenomena on the left side of the body, including the face. Thus, such sensations may be characteristic of trochlear nerve neurinoma and useful in the differential diagnosis of trochlear nerve neurinoma and trigeminal neurinoma (however, the neuroradiological findings are quite distinct). It is well known that the symptomatology of some neurinomas does not correspond to the nerves of their origin but rather to the functions of other, adjacent cranial nerves. For example, some trigeminal neurinomas manifest as abducens nerve palsy without trigeminal nerve symptoms.⁹⁾ Jefferson⁴⁾ explained this by noting that a neurinoma involves only part of the nerve fibers, and the tumor does not affect the root of the nerve. This seems to be true also of trochlear nerve neurinomas.

Precontrast CT scans demonstrated isodensity (Cases 4 and 5) or slightly high density (Case 6) with positive contrast enhancement in all four cases in which CT was performed. In our case the low-density mass seen on precontrast CT was not clearly enhanced after the administration of contrast medium. This finding led us to the preoperative misdiagnosis of brain stem glioma (if intra-axial) or epidermoid tumor (if extra-axial).

We feel that a trochlear nerve neurinoma is best

Fig. 4 Photomicrographs showing the features characteristic of neurinoma. *left*: $\times 100$, *right*: $\times 400$.

Table 1 Summary of reported cases of trochlear nerve neurinomas

Case No.	Author (Year)	Age (years) and sex	Duration of symptoms	Clinical manifestation	CT findings	Approach and location of tumor	Histology
1	King (1976) ⁵⁾	55F	1 year	gait disturbance, "pins and needles" sensation of the face, hemisensory disturbance		subtemporal; incisura	Antoni type A and B
2	Boggan <i>et al.</i> (1979) ¹⁾	32F	2 years	fourth nerve palsy, "tingling" sensation of the face, hemiparesis	precontrast: ?; enhancement	subtemporal; incisura	Antoni type A and B
3	Ho (1981) ³⁾	58F	incidental findings	none		autopsy; dorsal to the midbrain	Antoni type A and B
4	Leunda <i>et al.</i> (1982) ⁶⁾	54M	7 months	headache, diplopia	precontrast: isodensity; marked enhancement	subtemporal, transtentorial; incisura	schwannoma
5	"	16F	5 months	headache, diplopia	precontrast: isodensity; enhancement	subtemporal, transtentorial; incisura	schwannoma
6	Yamamoto <i>et al.</i> (1984) ⁸⁾	37F	1 month	headache, diplopia	precontrast: slightly high density; marginal enhancement	subtemporal, transtentorial; incisura	Antoni type A
7	Present case	43M	1 year	gait disturbance, "queer" sensation of the face, hemisensory disturbance	precontrast: low density; no enhancement	subtemporal, transtentorial; incisura	Antoni type A and B

approached through the subtemporal transtentorial route. These tumors are usually situated under the tentorium, compressing the midbrain medially, and are sometimes attached to the tentorial incisura. In six of the reported cases the trochlear nerve was identified in the posterior or posteroinferior aspect of the tumor, far from its exit from the midbrain. The tumor found incidentally at autopsy was situated about 1.5 mm from the trochlear nerve exit, that is, at the dorsal surface of the midbrain.

All six operated patients developed trochlear nerve palsy after surgery. Evidently, it is difficult to remove these tumors without compromising the trochlear nerve function.

In summary, physicians should be aware that the symptomatology of neurinomas does not always reflect the function of the nerve that is involved. It is to be noted that unusual facial sensations accompanied by contralateral hemiparesis and unilateral sensory disturbance even without trochlear nerve palsy may well be an initial manifestation of a trochlear nerve neurinoma.

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