SHORT REPORT

Cisternal trochlear nerve schwannoma: improvement of diplopia after subtotal tumour excision

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Abstract
Objective and importance. Trochlear nerve schwannomas without neurofibromatosis are very rare. No more than 33 cases have been pathologically verified. None of the reported cases showed postoperative improvement of trochlear palsy. The authors present a case of trochlear palsy due to a trochlear nerve schwannoma with subsequent improvement of trochlear palsy after subtotal excision of the tumour. Clinical presentation. A 65-year-old woman presented with diplopia, vertigo and left arm weakness. There was no history of neurofibromatosis. MR imaging demonstrated a tumour in the right perimesencephalic/ambient cistern. Intervention. The tumour was operated by a pterional approach, and subtotal excision was achieved preserving the tumour capsule with the trochlear nerve. The tumour was pathologically diagnosed as a schwannoma. Conclusion. At 2-year follow-up, there was no double vision and MR imaging showed a small, stable tumour remnant. To our knowledge, this is the first reported case of a trochlear schwannoma in whom postoperative improvement of trochlear function was achieved.

Keywords: cranial nerve; pterional approach; schwannoma; trochlear nerve

Introduction
Schwannomas are slow-growing, benign tumours that arise from Schwann cell populations at the central and peripheral myelin nerve junction.1 Sixty percent of all schwannomas arise from the cranial nerves (CNs). Schwannomas show a predilection to sensory CNs. Only rarely have they been reported to originate from motor CNs, frequently associated with neurofibromatosis. Overall, trigeminal nerve schwannomas are the most common non-vestibular CN schwannomas, followed by glossopharyngeal, vagal, facial, accessory, hypoglossal, oculomotor, trochlear and abducens nerve schwannomas. The problem with trochlear schwannomas is that preservation of the trochlear nerve after tumour removal is an extraordinary challenge. Furthermore, the chance for postoperative improvement of trochlear nerve function is quite low. Here, we report the unique case of a patient who achieved normalisation of trochlear nerve function after subtotal tumour removal.

Case report
History and examination
A 65-year-old woman was referred with a history of dizziness, vertigo and diplopia. Neurological examination revealed double vision on downward gaze, indicating trochlear nerve palsy. There was weakness and hypaesthesia in the left upper limb. There were no other focal neurological signs. No cutaneous signs of neurofibromatosis were evident. MRI demonstrated a well-defined tumour (diameter: 1.7 cm) located in the rostral right ambient cistern compressing the right cerebral peduncle (Fig. 1a, b). The mass extended anteriorly to the cavernous sinus and displaced the posterior cerebral artery posteriorly. The lesion was hypointense on T1-weighted and hyperintense on T2-weighted images with homogenous contrast enhancement.

After discussion with the patient, surgery was recommended and accepted with the aim of relieving the symptoms of brainstem compression. Conservative management was not desirable because of progressive arm weakness.

Operative procedure
Surgery was performed under general endotracheal anaesthesia. The patient was placed in a supine position with the head rotated 45 degrees to the left side and fixed in a Mayfield clamp. A typical right pterional craniotomy was made. After dural opening, CSF was drained by opening the Sylvian fissure and the optico-carotid cistern, followed by relaxation of the brain. Neurovascular structures were dissected and preserved. The Sylvian fissure was opened further to visualise the tentorial edge where the tumour came into view. After opening of Liliquist’s membrane, the basilar artery and the posterior cerebral artery were dissected. The tumour was found to be adherent to the tentorium. The trochlear nerve could not be dissected from the tumour capsule. On further dissection between the basilar artery and the tumour, the abducens nerve was visualised. Then the tumour capsule was opened, followed by central stepwise debulking. After incision of the
It arises from the midbrain below the inferior colliculi and passes around the brainstem near the junction of the midbrain and pons, to reach the lower margin of the free tentorial edge. The trochlear nerve pierces the medial edge of the tentorium and after a short distance in the anterior petroclinoid fold, it enters the lateral wall of the cavernous sinus.

Trochlear schwannomas are classified into three anatomical subtypes – cisternal, cisternocavernous and cavernous – according to their location and their relationship to the nerve segments.

Previously reported cases showed variable clinical presentations, such as features of other CN affection, in particular trigeminal signs, headache, long tract motor or sensory signs, cerebellar manifestations, e.g. ataxia and dysmetria, atypical facial pain and pathological laughter. Trochlear nerve palsy was evident in 44% of patients.

A preoperative definitive diagnosis based on the clinical picture and neuroimaging may be difficult, but trochlear schwannoma should be considered if there is one or more of the clinical signs outlined above with a mass in the ambient cistern, ventrolateral to the midbrain and pons, just beneath the tentorial edge.

In a series of 30 presumed trochlear nerve schwannomas, Elmalem et al. found that the majority of these patients tolerated their fourth nerve palsy very well without any treatment at all. In their series, the majority of the patients could be managed conservatively by follow-up MRI scans and many of them did not even require strabismus surgery.

Stereotactic radiosurgery (SRS) has shown high success rates in controlling schwannomas and should be taken into consideration as an alternative, minimally invasive technique.
especially in older patients with small tumours and in patients with higher risk for general anaesthesia due to additional diseases. Nevertheless, in tumours with local signs of brainstem compression, as in our case causing left arm weakness, surgery should be considered as the first treatment option. Also, when MRI scans are doubtful with regard to the diagnosis, surgery is an option to ascertain histopathological diagnosis.

The traditional approach to non-vestibular CN schwannomas has been surgical resection. In contemporary surgery, the combination of skull base approaches and microsurgical techniques provides satisfactory results. Different approaches have been reported for surgical excision of trochlear schwannomas: subtemporal transtentorial (12 cases), lateral suboccipital (6 cases), petrosal (anterior, posterior and combined) (5 cases) and pterional (4 cases).

In the previously reported cases, 12 patients had a preoperative trochlear palsy that did not improve postoperatively, while 17 cases had no preoperative trochlear palsy, and 12 of those developed a new postoperative trochlear palsy. This exemplifies the difficulty in preserving the trochlear nerve during trochlear schwannoma surgery and its vulnerability.

In our case, we used the traditional pterional transylvian approach, which is fast, straightforward and versatile. The wide range of neurovascular structures within reach, in addition to the variable working angles and the surgical freedom to manipulate instruments make it the standard approach to the middle fossa, and perimesencephalic region. Subtotal tumour extirpation was performed to offer the chance of improvement of IV nerve function.

Although a trochlear nerve deficit may be well compensated, restoration of normal function is a goal that might be achieved. Although the majority of patients tolerate well a fourth nerve palsy, we think that subtotal tumour removal preserving the tumour capsule with the trochlear nerve can be an appropriate technique in selected patients.

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References