An unusual presentation of neurofibroma of the oculomotor nerve

Case report

PAUL Hiscott, M.B., B.S., AND LINDSAY Symon, F.R.C.S.

Department of Neurological Surgery, National Hospital, Queen Square, London, England

The authors describe a patient who presented with progressive dementia and hemiparesis. A neurofibroma was demonstrated arising from the intracavernous portion of the contralateral oculomotor nerve and the patient was treated successfully by subtotal excision.

KEY WORDS • oculomotor nerve • neurofibroma • neurilemmoma

NEUROFIBROMAS of the oculomotor nerve are very rare, and may be seen in multiple neurofibromatosis or as isolated lesions. They usually present with symptoms and signs of third cranial nerve damage and, if they arise within the cavernous sinus, also with the features relating to involvement of other intracavernous structures (“cavernous sinus syndrome”). It is distinctly unusual for such an isolated lesion to produce hydrocephalus and hemiparesis with only minimal third-nerve signs. Recently, we treated such a case.

Case Report

This 58-year-old woman presented to a general hospital with a 6-month history of gradual deterioration in memory, with increased lability of mood, and a slight paresis of the right leg. Six weeks before admission, her relatives had noted the onset of mild but increasing confusion, associated with urinary incontinence. At about that time, the paresis began to involve the right arm and side of the face. The patient herself complained of minimal intermittent frontal headache and drowsiness, not related to any time of the day. The medical history was unremarkable except for long-standing right-sided deafness. On admission, she had mild bilateral papilledema, in addition to the mild dementia and hemiparesis. The patient was started on a course of dexamethasone, 16 mg daily, and subsequently became more alert. She was transferred to the neurosurgical unit for investigation.

Examination. The patient was apyrexial and slightly drowsy but cooperative, with a minimal expressive dysphasia. Arithmetic function and short-term memory were grossly impaired. Her stance was unsteady and she had a positive Romberg's sign; there was evidence of a hemiparetic gait. Ophthalmologically, visual acuity was correctable to 6/6 (and N5 reading) in each eye. There was chronic papilledema of the left optic disc, with less marked papilledema on the right side. Formal visual field analysis revealed no objective defect, although subjectively there may have been a difference between the right and left half-fields. Apart from a possible transient left afferent pupillary defect, pupillary responses were normal. There was mild ptosis of about 1 mm of the left upper eyelid, but no other evidence of third nerve damage, and no extraocular muscle paresis. An upper motor neuron facial weakness was noted on the right side, and a conductive deafness on the same side was confirmed. Other cranial nerves, including the trigeminal nerves, were normal. There was a mild right-sided motor weakness, power being 4/5 compared to 5/5 on the left (Medical Research Council recommended grading). There was no alteration in tone, and all reflexes were equal with flexor plantar responses. No sensory abnormalities were elicited. On general examination, the pulse was 100 beats/min and blood pressure 110/80 mm Hg. There were neither café-au-lait spots nor other evidence of von Recklinghausen's disease.

Routine hematology, biochemistry, and skull and chest radiology were normal. Computerized tomog-
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FIG. 1. Computerized tomography scan, axial view, with injection of contrast material, showing enhancing tumor and hydrocephalus.

raphy revealed an enhancing lesion adjacent to the inner third of the sphenoid ridge, associated with hydrocephalus (Fig. 1). Left carotid angiography demonstrated pathological circulation and confirmed the size of the lesion. Meningioma was thought to be the most likely diagnosis.

Operation. At left frontotemporal craniotomy (L.S.), a large mass with a smooth surface was found extending into the inner end of the Sylvian fissure. Dissection revealed that the tumor occupied and erupted from the cavernous sinus. It extended to the interpeduncular cistern. It had the macroscopic appearance of neurofibroma, but the fifth nerve was stretched below it and was not involved in the tumor. The third nerve, however, appeared from the interpeduncular cistern as a stout bundle, and ran straight into the mass, spread out, and disappeared. The tumor was excised except for a small residuum closely adherent to the carotid artery. The left oculomotor nerve was the only structure giving origin to it. The fourth cranial nerve was visualized and appeared intact outside the cavernous sinus.

Histological Study. Macroscopically, the tumor was a gray, friable lesion. Microscopy revealed a benign neurofibroma consisting of intertwining bundles of elongated cells with rod-shaped nuclei intermingled with looser areas of smaller cells with round or oval nuclei. The latter had more abundant collagen (Fig. 2).

Postoperative Course. After surgery, the patient showed a complete third and partial fourth cranial nerve palsy on the left. The hemiparesis completely resolved. Her initial progress was delayed by a mild reactionary hemorrhage and exacerbation of the hydrocephalus, requiring a ventriculoperitoneal shunt. She also suffered a deep venous thrombosis and pulmonary embolism, which responded to anticoagulant therapy. The patient exhibited withdrawal behavior with anger outbursts, for which antidepressants were given with good response. Four months postoperatively, the troclear nerve paresis and psychic disturbances had completely resolved; the only remaining neurological deficit was the complete oculomotor nerve palsy.

Discussion

The terms “neurilemmoma,” “neuroma,” “neurilemmoma,” and even “neurofibroma” have been used for Schwann cell tumors, although “neurofibroma” has usually been applied to tumors arising from fibroblasts. Recently, evidence has been presented that even tumors classified as neurofibromas derive from Schwann cells. Histologically, the tumor in our report had features of both schwannoma and neurofibroma.

Neurofibromas represent no more than 9.3%, and perhaps less than 8%, of all intracranial neoplasms. The vast majority arise on sensory nerves, most commonly the eighth, and then the fifth cranial nerves. Involvement of other cranial and especially motor nerves is very rare in patients without von Recklinghausen’s disease. Any cranial nerve may be involved in multiple neurofibromas. Bailey, et al., and Ford are among those who have reported oculomotor nerve involvement in this disease. Isolated neurinomas of the third nerve are very rare.

In 1927, Kovacs demonstrated an isolated third-nerve neurinoma at autopsy. In 1975, Shuangshoti reported a case with a third-nerve palsy. Huber presented three patients who had symptoms and signs of oculomotor nerve paresis, preceded or paralleled by functional loss of the ipsilateral optic nerve. Schubiger, et al., described a neuroma of the cavernous sinus which may have arisen from the oculomotor nerve, with third, fourth, and fifth nerve involvement. Such a “cavernous sinus syndrome” is also associated with other lesions in the cavernous sinus, including aneurysms and tumors. A lesion involving the oculomotor nerve may be expected to cause symptoms...
and signs of third-nerve paresis,\textsuperscript{3,10,14} and, if arising in the cavernous sinus, visual impairment and pain in the trigeminal distribution (upper two divisions) in addition. Our patient did not have von Recklinghausen’s disease, in keeping with most reports, but further presented with mild dementia and contralateral hemiparesis. There were minimal second- and third-nerve signs. The dementia was presumably due to the secondary hydrocephalus, and the hemiparesis to pressure on the upper brain stem at the tentorial level.

Fibers on the surface of the third nerve supply the levator palpebrae muscle, and hence pathology which involves these fibers may cause ptosis.\textsuperscript{6} Our patient had minimal ptosis, and perhaps only those superficial fibers were compromised by the tumor. The transient afferent pupillary defect may have been caused by disc edema secondary to elevated intracranial pressure, or due to direct pressure on the optic nerve by the tumor. Interestingly, it has been noted that a fourth-nerve neuroma may not necessarily present with diplopia,\textsuperscript{6} although, as with a third-nerve tumor, such may occur at an early stage.\textsuperscript{2}

Both Boggan, \textit{et al.},\textsuperscript{2} and Schubiger, \textit{et al.},\textsuperscript{11} point out that neuromas of the oculomotor and trochlear nerves may be misdiagnosed as trigeminal tumors at surgery. Preoperatively, the most likely diagnosis in our patient was considered to be meningioma of the sphenoid ridge, as in Huber’s three patients.\textsuperscript{4} The operative findings and postoperative course left no doubt as to the diagnosis.

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\textbf{References}


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\textsuperscript{Address reprint requests to: Professor Lindsay Symon, Institute of Neurology, Queen Square, London WC1N 3BG, England.}