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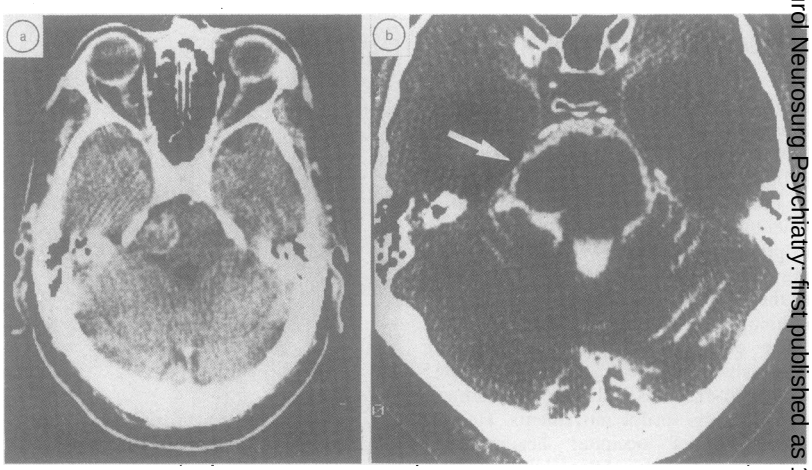

Fig a *CT: after contrast enhancement. Shows oval well-defined lesion in the side of the brain-stem.* b *CT: with intrathecal contrast. Tumour (arrowed) is shown to be extrinsic to the brain-stem.*

Isolated schwannoma of the fourth cranial nerve: case report

Sir: Schwannomas of the nerves supplying the external ocular muscles are exceptionally rare. The literature contains only six reports of such a tumour arising from the trochlear nerve. A further case is reported here.

A 56 year old van driver was referred with a five month history of increasing weakness and clumsiness of his right limbs together with slurring of speech. For two weeks he had suffered from morning headaches and double vision on gaze to the right. Before the onset of his first symptoms he had been in good health. There was no family history of neurological disease.

Non neurological examination was unremarkable; he showed no features of neurofibromatosis. His mental state and optic discs were normal. He had a marked spastic dysarthria. There were no abnormalities of the cranial nerves other than a right lower facial weakness. In particular, he had full external ocular movements without diplopia and no trigeminal deficit. He had a moderately severe spastic right hemiparesis in the limbs. The tendon reflexes were symmetrically brisk and both plantar reflexes were extensor. He was only just able to walk without assistance.

Skull radiographs showed no abnormality. Before enhancement the CT scan showed no abnormality. With contrast enhancement an oval irregularly hyperdense lesion 2cm in diameter could be seen, situated superficially in the left cerebral peduncle at the level of the tentorial edge (fig a). There was no distortion or displacement of the Aqueduct of Sylvius. CT after injection of intrathecal contrast showed that the mass was extrinsic rather than intrinsic (fig b).

The left cerebellar hemisphere was retracted via a left sided sub-occipital craniectomy, to permit inspection of the anterior part of the left cerebellopontine angle. A well-defined tumour 2 cm across was found partially embedded in the left side of the brain stem and lightly adherent to the back of the clivus at the level of the tentorium. The tumour lay above and medial to the left 7th and 8th cranial nerves and the left trigeminal sensory root was stretched over its upper pole. The far side of the tumour abutted the right trigeminal sensory root. The tumour was separated from the adjacent structures and totally removed. Removal involved the sacrifice of a single nerve of small diameter which blended into the tumour capsule. Histological examination revealed a typical schwannoma.

Post-operative recovery was uneventful. The patient complained of some inconstant double vision on gaze downwards and to the right with a tilted false image.

Two years later the patient had retired from work but was otherwise leading a full life with no disability. The patient showed the slight head tilt of a patient with a trochlear nerve palsy. His speech was now virtually normal and there was only a slight degree of spasticity of the right limbs. He was able to walk briskly but with a degree of evident stiffness of the right leg.

Most intracranial schwannomas arise from the 8th cranial nerve. Next in frequency, after acoustic schwannomas, are the trigeminal variety but they only account for 0·2% of intracranial tumours.[1] In the absence of von Recklinghausen's disease, schwannomas of the other cranial nerves are exceedingly rare. The first report of an isolated schwannoma of the 4th nerve was by King in 1976.[2] Including our patient, seven cases have now been reported.[2–6]

All but two of the tumours arose from the 4th nerve where it passes round the side of the cerebral peduncle, close to the tentorial edge. In one patient the origin of the tumour was probably more distal at or beyond the point where the nerve pierces the dura to enter the cavernous sinus, as the tumour lay mainly on the medial aspect of the middle fossa floor. Only one tumour was close to the point where the nerve becomes invested with schwann cells, a few millimetres after it has left the roof of the midbrain. Nevertheless, it is widely believed that the schwannomas of the cranial nerves tend to arise at the glial-schwann cell junction, as they appear to do in the case of acoustic tumours.[6]

All but one of the patients were aged between 32–58 years and five of the seven cases were women. The symptoms and signs resulted from a combination of dysfunction of the 4th nerve and compression of the adjacent cerebral peduncle and trigeminal sensory root. The tumour with a middle

fossa component involved the 3rd cranial nerve.

Plain radiographs were normal except in one case where there was erosion of the ipsilateral posterior clinoid process. As might be expected with a schwannoma, CT showed an isodense or slightly hyperdense mass with a well defined edge, which enhanced after intravenous contrast. When the tumour is embedded in the side of the brain stem at the level of the tentorial edge, it may be difficult to determine whether it is intrinsic or extrinsic until the scan has been repeated with intrathecal contrast, as in the present patient.

In all six patients who had surgery, a total removal was achieved with good functional recovery. In the case reported here, the tumour was removed from below via a suboccipital craniectomy. In the other patients the subtemporal transtentorial route was used.

It seems possible that schwannomas of the trochlear nerve and other nerves controlling external ocular movement may have been under-diagnosed in the past, being mistaken for trigeminal schwannomas. The curious fact that large "trigeminal" schwannomas may be associated with little or no disturbance of trigeminal nerve function is well known.[1] Before the invention of CT it was difficult to diagnose a tumour situated at the medial end of the petrous ridge before it became so large that it was difficult at the time of operation to determine with certainty the nerve of origin. Schwannomas of the oculomotor nerves arise close to the trigeminal sensory root and a large tumour, as in the second patient with a trochlear schwannoma described by Leunda *et al* may bridge the medial petrous ridge into the middle fossa like a trigeminal schwannoma.[4] Before operation the disturbance of external ocular movement may be minimal, as in the patient described in this report.

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An ependymoma involving the pituitary fossa

Sir: Ependymomas arising in the region of the third ventricle may occasionally cause visual loss by compressing the optic chiasm. Standard references of surgical neuropathology also refer to ependymomas in the differential diagnosis of intrasellar mass lesions, although we were unable to find any well documented cases. We therefore report a patient with clinical and radiological features of pituitary tumour which proved on transphenoidal removal to be an intrasellar ependymoma.

An 81 year old man had a six month history of progressive visual failure affecting particularly both temporal fields. There was a five month history of continuous headache felt over the right eye and three months of intermittent double vision with horizontal separation of images. His relatives had noticed a right ptosis. There was slight postural dizziness but no other symptoms to suggest endocrine dysfunction. Examination revealed visual acuities of 6/18 and 3/60 in the left and right eyes respectively, with a bitemporal hemianopia. There was a complete right third nerve palsy with preserved fourth and fifth nerve function. Blood pressure was 100/70 without postural drop.

He had a raised serum prolactin at 536 mu/ml (normal range 3–178 mu/ml) and a reduced free T4 (7 pm/l n > 8·8). CT scan (fig 1) showed a large enhancing mass lesion arising in the pituitary fossa and extending out of the sella, which was considered typical of a pituitary adenoma.

Because of the severity of the symptoms and the progressive visual failure, transphenoidal removal of the intrasellar tumour was carried out. The floor of the fossa was partially deficient and beneath a thin rim of pituitary the tumour was located and evacuated from the sella and suprasellar regions. Operation was uneventful, with no CSF leak noted, and was followed by immediate improvement in visual acuity in the right eye to 6/36. A few days following the operation he developed a fever and increasing confusion. Cerebrospinal fluid (CSF) removed at lumbar puncture contained 960 white cells/ml, with a protein of

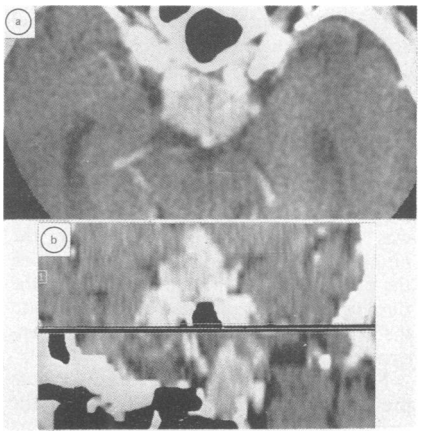

Fig 1 *(a) A high definition CT scan in the immediate suprasellar region; (b) computerised coronal and saggital showing a continuous intra-suprasellar lesion.*

580 mg% and 0·6 mm/l of sugar. Microscopy showed numerous gram positive rods and cocci. Despite treatment with intravenous cloramphenicol, metronidazole, penicillin and steroids he rapidly deteriorated and died three days later. Permission for post mortem was refused.

The pathological specimen consisted of several small fragments of grey tissue with a gelatinous consistency in some parts. Examination of paraffin sections stained for routine histology (fig 2) showed a tumour made up of elongated cells arranged in loosely packed rows and perivascular pseudorosettes. The tumour cells had eccentric pleomorphic nuclei, coarse chromatin and there were moderate numbers of mitotic figures. Glial fibrillary acidic protein, a marker for astrocytic-ependymal differentiation, was present in the processes of the tumour cells. This appearance is diagnostic of a moderately malignant ependymoma.

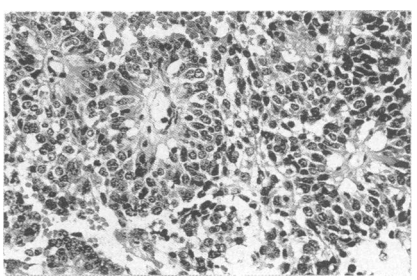

Fig 2 *Photomicrograph showing tumour cells arranged to form perivascular rosettes around thin capillaries (haematoxylin and eosin *300).*