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Intracranial extension of a naso-ethmoid schwannoma

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Abstract
A rare case of intracranial extension of a naso-ethmoid schwannoma is presented. Its subsequent removal with few sequelae confirmed the benign nature and good prognosis of the disease.

Introduction

Schwannomas of the naso-ethmoid region are uncommon, approximately 35 cases having been reported in the world literature (Khalfifa and Bassyouni, 1981). Intracranial extension has been reported once (Zovickian *et al.*, 1986). We report a tumour originating in the left ethmoid complex, with massive intracranial extension. We outline its management, clinical and pathological features, and discuss possible anatomical sites of origin.

Case report

A 28-year-old Caucasian male presented with a nine month history of recurring headaches associated with nausea and vomiting. This was preceded by a severe pain at the root of his nose which lasted for several days and which settled spontaneously. The headaches became progressively worse, and three months prior to neurosurgical consultation he had absence attacks and intermittent blurred vision. According to his relatives, his personality had become more out-going and disinhibited over this time. On examination, he was slightly obese, well-looking and cheerful. He had bilateral papilloedema, hyposmia on the left and brisk tendon reflexes in his upper limbs. Examination was otherwise normal.

Computed tomographic scans (Figs.1–3) showed a large

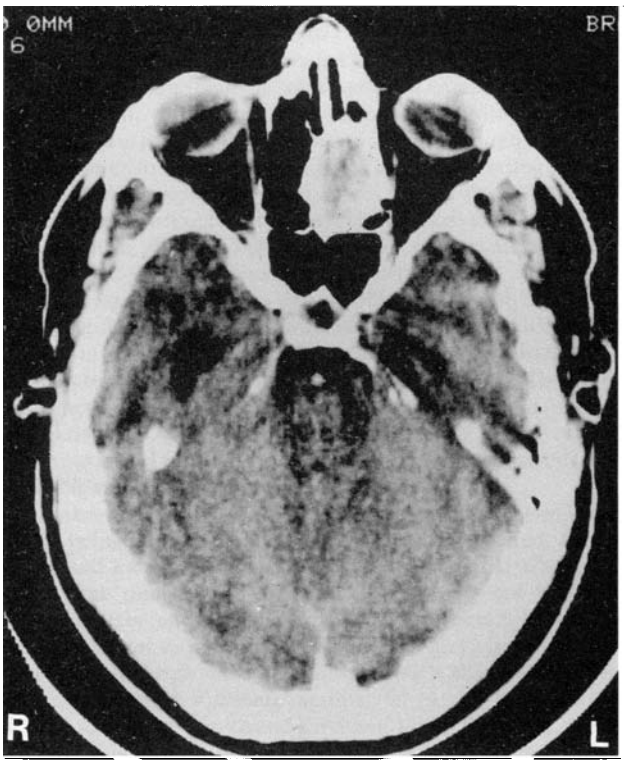

Fig. 1
Tranverse section of skull showing tumour in left ethmoid sinus.

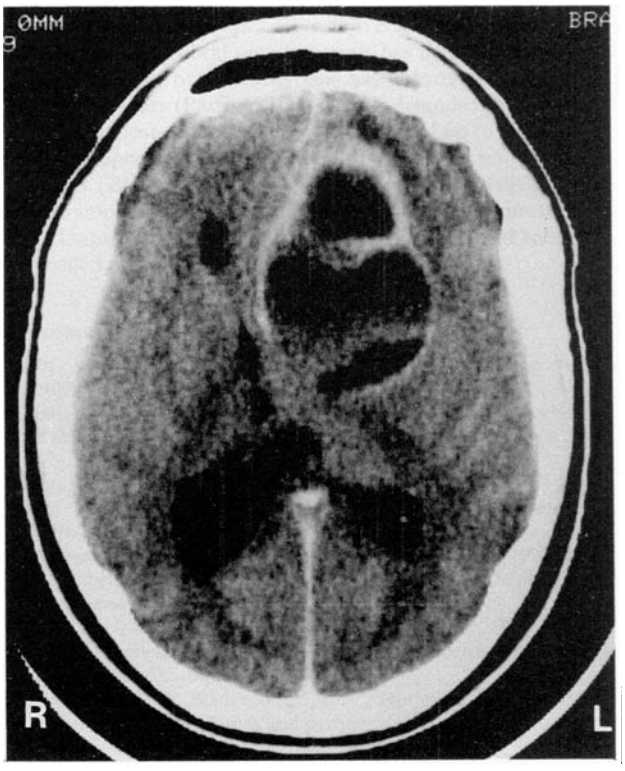

Fig. 2
Tranverse section of skull showing tumour in left frontal lobe with gross distortion of ventricular system.

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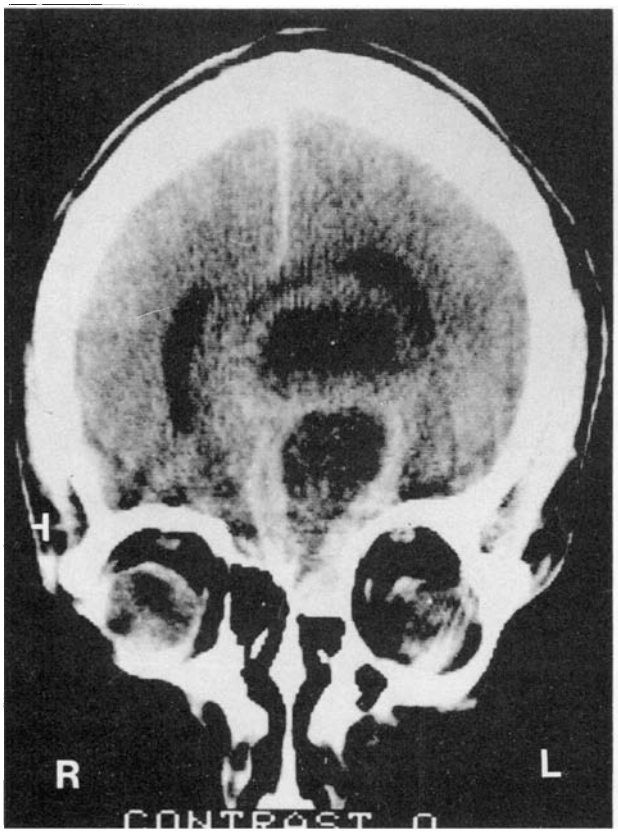

FIG. 3

Coronal section through skull showing tumour spreading from left ethmoid sinus through cribiform plate and into brain.

mid-line frontal mass extending to the left, with a maximum diameter of 7 cm and involving the left ethmoid complex. There was compression of both frontal lobes and distortion of both lateral ventricles. It was of low attenuation but the rim enhanced with intravenous contrast medium.

The day following admission, a bifrontal craniotomy was undertaken. The left frontal lobe was elevated to display a smooth white vascular tumour. It was easily separated from the surrounding brain. Internal decompression was facilitated by the drainage of two large cysts filled with dark-coloured fluid suggesting degeneration and haemorrhage. The remainder of the inside of the mass was firm but degenerate and was easily removed by suction and curettage. The firm outer part of the tumour was removed easily, the only attachment being in the region of the cribiform plate (Fig. 4). Dissection in this area revealed a dumb-bell extension through the dura into the left ethmoid sinus which was entirely filled with tumour. This was easily removed from above and the dural defect was repaired with a graft of lyophilized dura.

Post-operatively, there was a minor cerebrospinal fluid rhinorrhoea which settled spontaneously after three days. Disinhibition was increased for a few weeks, but when seen three months later he was thought by his family to be back to normal and there were no abnormal neurological signs, including his olfactory sensation.

Histological examination showed the tumour to be composed of compact interlacing bundles of elongated cells showing a tendency to form nuclear 'palisades' (Fig. 4). There were also scattered collections of cells with distended foamy ('xanthomatous') cytoplasm (Fig. 5). There was abundant reticulin fibre production interspersed between cells. The tumour did not contain admixed nerve fibres, but stainable axons were demonstrated in the periphery of the tumour. Mitotic figures were not seen. The appearances were those of a schwannoma.

Discussion

Schwannoma may arise on any nerve with a schwann cell

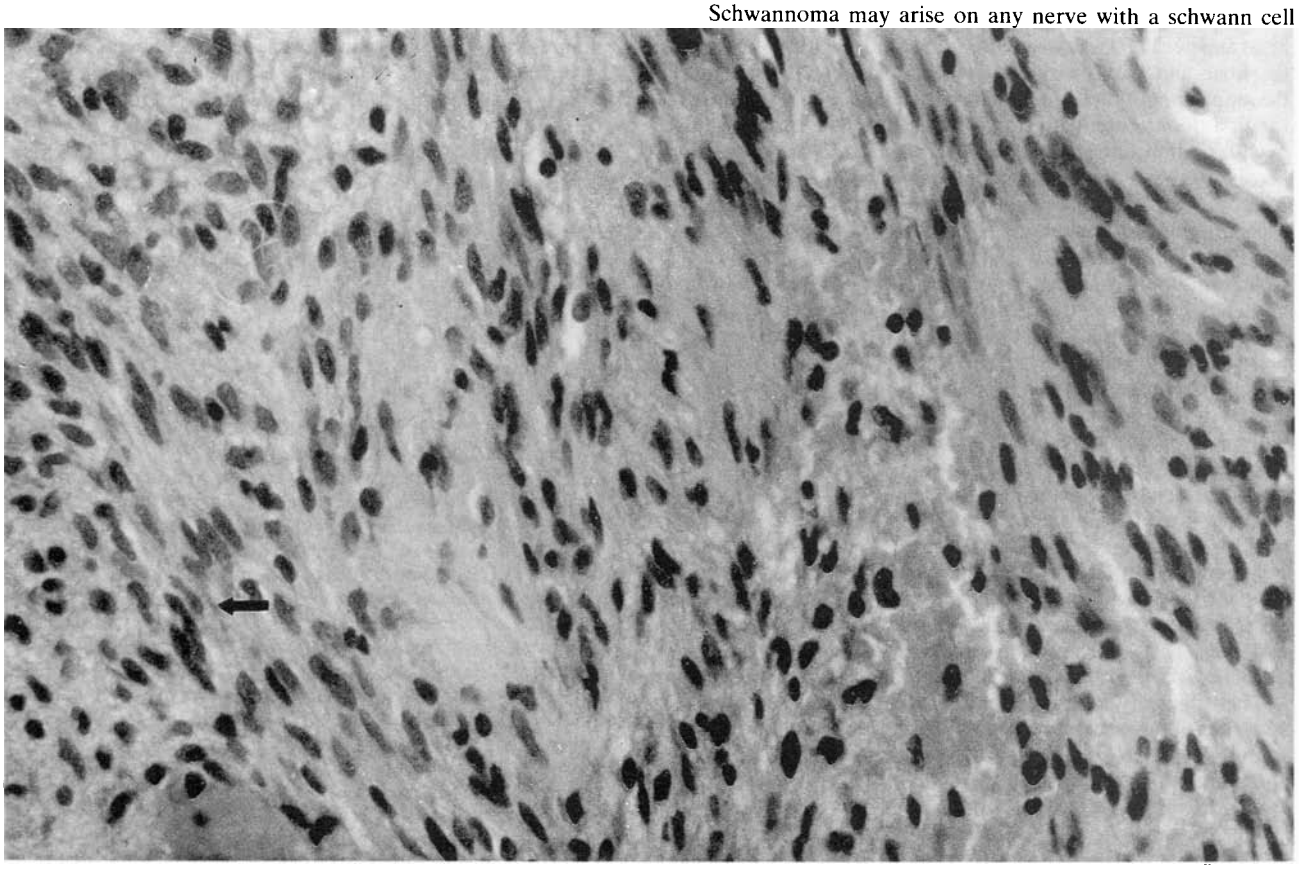

FIG. 4

Photomicrograph showing interlacing bundles of elongated cells and characteristic nuclear 'palisades' (arrow). Haematoxylin and eosin (H&E ×500).

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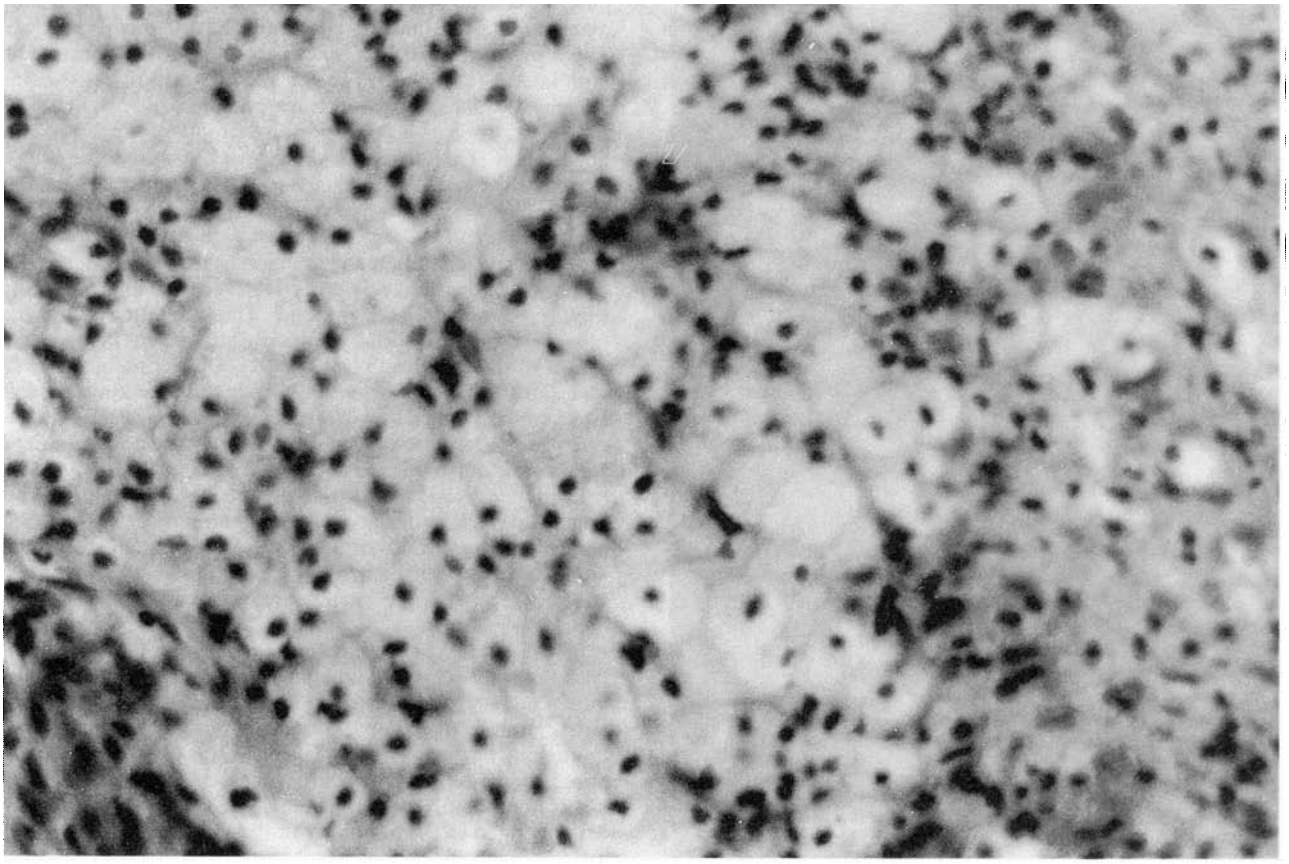

FIG. 5

Photomicrograph showing collections of 'xanthomatous' cells. (H&E ×500).

sheath (Batsakis, 1979). Up to 45 per cent of them occur in the head and neck (Hawkins and Luxford, 1980). Schwannomas in the nose and para-nasal sinuses, however, are unusual. Of these, naso-ethmoid tumours are the most common (this was the first documented site) followed by tumours arising in the maxillary sinus, intranasally and in the sphenoid sinus (Baksakis, 1979). None have been found in the frontal sinuses (Calceterra *et al.*, 1974). There is no association with race or sex. Most present between the ages of 20 and 60 years (Calceterra *et al.*, 1974; Robitaille *et al.*, 1975).

Naso-ethmoid tumours commonly spread locally causing nasal blockage and epistaxis. In this case the slowly progressive growth through the roof of the ethmoid sinus and into the cranial cavity allowed the tumour to reach a considerable size without obvious symptoms, in the same way as primary intracranial tumours of the frontal region (Batsakis, 1979). Surprisingly, in view of the size of the tumour, nasal symptoms were minimal. Olfactory sensation was preserved pre- and post-operatively.

These tumours can be highly vascular and severe bleeding may be provoked by biopsy (Harkins, 1949). Despite this, enlargement can lead to areas of cystic degeneration as this supply is outgrown, as our case showed. The cystic degeneration facilitated removal by allowing drainage of the tumour contents. This kept surgical trauma to a minimum, and so reduced risks of neurological sequelae.

The origin of this tumour is debatable, as there are many branches of the ophthalmic and maxillary nerves which ramify in this vicinity. We believe it is most likely to have arisen from the ethmoid branches of the nasociliary nerve. It may, however, be from an autonomic source originating in the pterygopalantine ganglion, which supplies the area in question via its orbital branches (Williams and Warwick, 1980). The olfactory nerves are covered by glial cells and so cannot give rise to nerve sheath tumours (Batsakis, 1979); they were found to be separate from the tumour at operation.

There was no evidence of other neurofibromas in this patient. Naso-ethmoid schwannomas have not been reported in association with von Recklinghausen's disease (Gignoux and Labayle, 1949), and all other tumours so far reported have been solitary (Harkins, 1949; Hawkins and Luxford, 1980; Iwamura *et al.*, 1972; Khalfifa and Bassyouni, 1981; Zovickian *et al.*, 1986).

The best treatment for schwannomas remains complete surgical resection (Shugar *et al.*, 1982). The prognosis is excellent, since the vast majority are benign (Robitaille *et al.*, 1975).

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