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An extensive vestibular schwannoma with both intracranial spread and lateral extension to the external auditory canal

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Abstract
An extensive schwannoma in a 54-year-old woman is described. There was extension intracranially to involve the brain stem, laterally to fill the middle ear and external meatus, and inferiorly into the infratemporal fossa. Initially the intracranial portion of the tumour was excised. Later a second operation was undertaken to remove the residual tumour from the temporal bone and infratemporal fossa.

Key words: Schwannoma, vestibular; Ear, external; Ear, middle

Introduction

Schwannomas of the VIIIth cranial nerve are benign tumours which usually arise from the superior or inferior vestibular nerves within the internal auditory meatus (IAM) and grow medially taking the line of least resistance. They account for approximately three-quarters of cerebellopontine angle (CPA) tumours. Lateral extension of a vestibular schwannoma with involvement of the middle or external ear is very uncommon as is extension into the infratemporal fossa. In this paper we report the case of a large schwannoma which extended from the cerebellum medially, through the temporal bone to present laterally in the external auditory meatus, and inferiorly into the infratemporal fossa.

Case report

A 54-year-old lady was referred to the neurosurgical department of another hospital with a 20-year history of left-sided audiovestibular symptoms which began with a sudden sensory neural hearing loss. Her hearing continued to deteriorate, and the initial vague imbalance eventually became severe. Examination suggested a dead ear on the left (which was confirmed by audiometry), an incomplete left lower motor neurone facial palsy, reduced sensation over the distribution of the trigeminal nerve and early palatal weakness. Romberg's sign was positive and there was evidence of decreased cerebellar function on the left side. CT (Figure 1) and MR imaging revealed a CPA tumour encroaching on the brain stem medially and extending laterally into the petrous temporal bone. There was no hydrocephalus. The patient underwent excision of the intracranial portion of the tumour via a retrosigmoid craniectomy but no attempt was made to pursue the tumour into the internal auditory meatus or petrous bone. Histological examination confirmed a schwannoma with both Antoni A and B areas. The patient made a good recovery from the surgery although her facial nerve palsy increased.

Two years after the original surgery she was referred to the Manchester Royal Infirmary Otolaryngology Department with increased difficulty in swallowing. Examination revealed a tumour in the left external auditory canal (Figure 2), a complete lower motor neurone facial nerve palsy, decreased trigeminal nerve function and palsies of the lower cranial nerves indicated by palatal weakness, a compensated vocal fold palsy, atrophy of sternomastoid and wasting and fibrillation of the tongue. CT and MR imaging (Figure 3) demonstrated a tumour occupying the whole of the left petrous temporal bone, extending into the ear canal and inferiorly into the neck.

Biopsy via the ear canal confirmed that the tumour was a

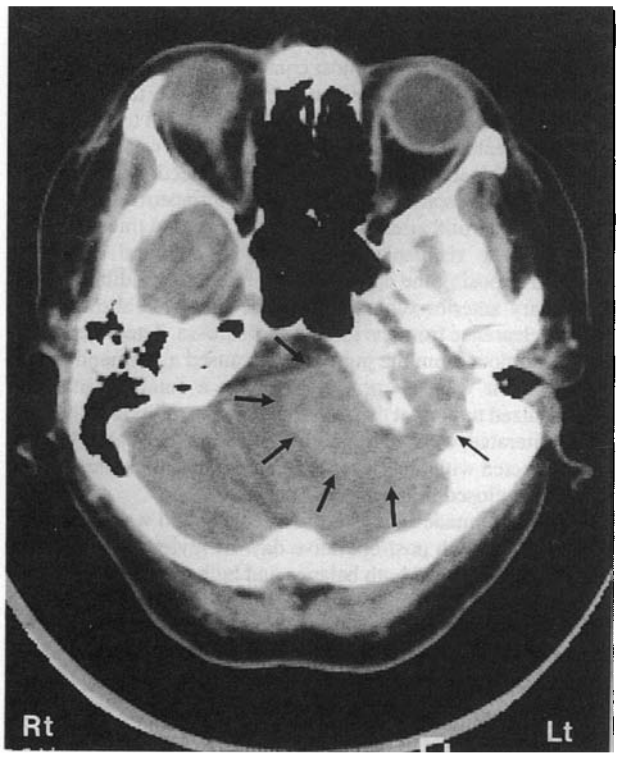

FIG. 1

Original CT scan prior to first operation showing large left cerebellopontine angle (CPA) lesion (arrowed) crossing the midline and causing extensive temporal bone destruction.

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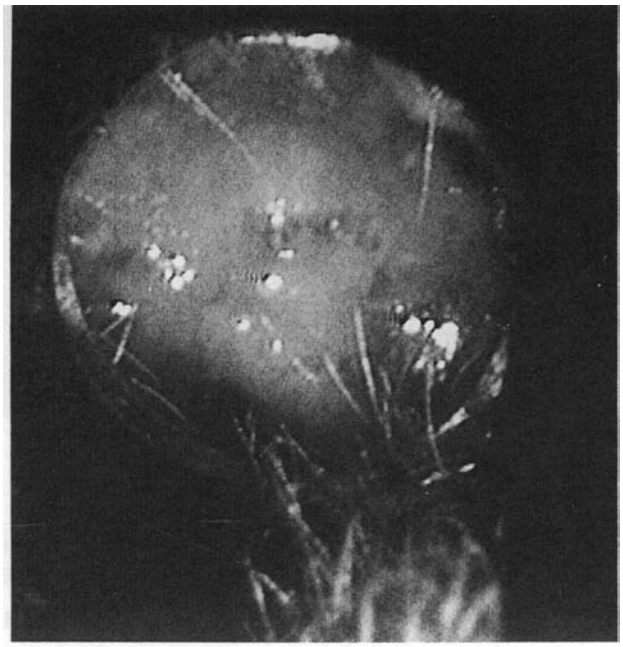

FIG. 2
Schwannoma visible in the external auditory canal.

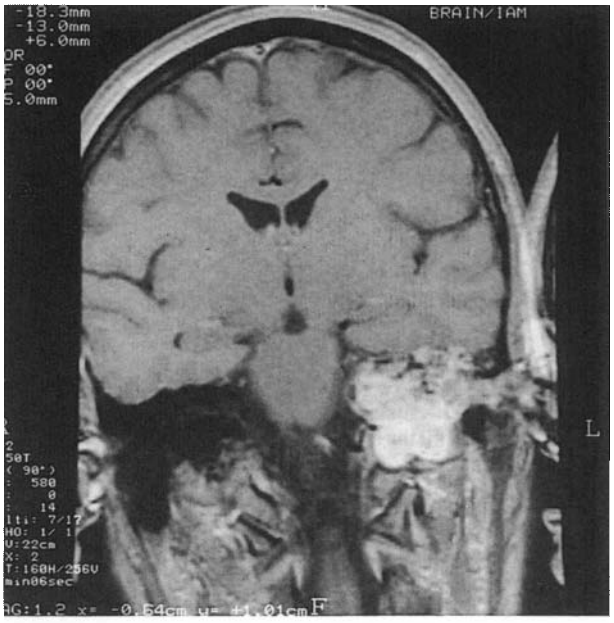

FIG. 3
MR imaging prior to second operation. Tumour is seen to fill the temporal bone and to extend into the external auditory canal. There is destruction of the skull base in the region of the jugular foramen with extension of tumour into the infratemporal fossa.

schwannoma and the patient subsequently underwent exploration of the left temporal bone, posterior cranial fossa and infratemporal fossa via a post-auricular approach with the incision extended down the anterior border of the sternomastoid. Tumour was found to fill the whole of the temporal bone, eroding through the mastoid cortex to lie subcutaneously behind the ear. Widespread exposure of the posterior and middle fossa dura was performed. There had been almost complete destruction of middle ear structures, facial nerve and cochlea by the tumour which extended from the ear canal to a point just medial to the internal auditory meatus. Extensive tumour excision was undertaken using a combination of diathermy, sharp dissection and continuous ultrasound aspiration (CUSA). The intrapetrous portion of the internal carotid artery was dissected free and traced into the neck and the tympanic portion of the temporal bone was removed exposing the capsule of the temporomandibular joint. Temporary anterior subluxation of the mandible allowed total tumour clearance from the infratemporal fossa. Attempted clearance of tumour from the jugular bulb caused a profound bradycardia and it was felt prudent to leave a small fragment of diathermized tumour at this site. The cavity was lined with fascia and obliterated with abdominal fat, the internal carotid artery was protected with the digastric and sternomastoid muscles and the wound closed in layers.

The patient made an uneventful recovery and was discharged home on the ninth post-operative day. At review in outpatients she remains well and both balance and bulbar function continue to improve.

Discussion

Vestibular schwannomas typically arise from the vestibular nerve at the glial–neurilemmal junction usually located within the IAM but not uncommonly more medially in the posterior cranial fossa. They grow medially into the CPA and several clinical stages related to tumour size are recognized (Ramsden, 1987). Initially intrameatal and extrameatal tumours up to about 2 cm in diameter cause almost exclusively audiovestibular symptoms (the otological stage). Symptoms related to trigeminal nerve involvement follow and rarely a facial nerve palsy is seen with larger tumours.

Next there is evidence of cerebellar and brain stem compression with ataxia and lower cranial nerve dysfunction. A later stage still is characterized by headaches, vomiting and failing vision due to raised intracranial pressure preceding terminal disturbance of vital brain stem function. Vestibular schwannomas are now commonly diagnosed when 'otological' symptoms predominate, but review of this patient's original CT scans confirm that at the time of the first operation the intracranial tumour was large (4 cm).

Schwannomas arising other than in the IAM or posterior cranial fossa are rare. Thomsen and Jorgensen (1973) reported a case of an intracochlear neuroma which was seen to originate in the spiral ganglion, and Storrs (1974) described two cases of vestibular schwannoma presenting as middle ear tumours. Tran Ba Huy *et al.* (1987) described the only case of extension into the external auditory canal. De and Basu (1989) described a case of a vestibular schwannoma presenting as a subcutaneous swelling over the mastoid. Extension into the infratemporal fossa is very rare. Schwannomas arising within the vestibule have been reported (Birzgalis *et al.*, 1991) usually as a chance finding at labyrinthectomy carried out for the relief of undiagnosed vestibular symptoms.

The possible sites of origin of the tumour in this case are: (1) the internal meatus with medial and lateral extension; (2) the vestibule, with medial and lateral extension; (3) the facial nerve. This seems unlikely in view of the fact that there was little facial weakness before the original operation; (4) a multifocal origin with separate tumours arising in the internal meatus and in the temporal bone. Multifocal tumours may be seen in neurofibromatosis Type 2 (NF2) but there was no evidence of this condition either clinically or on MR imaging (Evans *et al.*, 1993) in this case.

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