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Clinical Records

Primary neuroblastoma of the facial nerve presenting as a recurrent facial paralysis

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Abstract
A case is described of a primary neuroblastoma of the facial nerve in a 13-year-old boy presenting with a recurrent facial paralysis. This tumour was excised preserving the nerve and followed with post-operative radiotherapy. The pathology of the tumour is described and facial nerve tumours briefly discussed as a cause of facial palsy. There have been no other cases of a primary neuroblastoma of the facial nerve arising at this site reported in the last 20 years.

Key words: Neuroblastoma; Facial nerve

Introduction

Facial palsy is a common clinical condition. Although idiopathic facial paralysis, or Bell's Palsy, is the commonest cause, this is a diagnosis of exclusion and must only be made after other aetiologies have been ruled out. In children, facial nerve paralysis is usually the result of infection, trauma or congenital disorder, and paralysis of neoplastic aetiology is rare. This is a reflection of the rarity of all types of head and neck tumours which, including metastases from distant primaries, account for five to 27 per cent of paediatric malignancies (Robinson *et al.*, 1988). Within this group primary neuroblastoma is particularly uncommon and in two large series, reporting on all head and neck malignancies in childhood, this tumour accounted for less than one per cent of cases (Jaffe and Jaffe, 1973; Robinson *et al.*, 1988). Primary neuroblastoma of the facial nerve is exceptionally rare and a thorough search of the literature in the English language has revealed no previous cases of a primary neuroblastoma at this site in the last 20 years.

Case report

A 13-year-old boy was referred to the ENT Department by his General Practitioner with a two-week history of right-sided facial weakness. There had been no preceding illness and, apart from the facial weakness, there were no other symptoms. He was fit and well in all other respects.

Examination revealed a right-sided lower motor neurone paralysis of the facial nerve, House Brackmann Grade 3. Other cranial nerves were intact and both tympanic membranes were normal to inspection, Rinne and Valsalva positive. Pure tone audiometry was normal in both ears, but stapedial reflexes were absent on the right side. He was treated conservatively and at review two weeks later full facial nerve function had been regained.

The patient was re-referred one month later with a recurrence of the palsy but on this occasion, in addition to the complete lower motor neurone paralysis, a red mass was visible behind the postero-superior quadrant of the ipsilateral tympanic membrane. CT scanning showed a soft tissue mass filling the epitympanum from the anterior attic to the aditus and surrounding the long process of the incus (Figure 1).

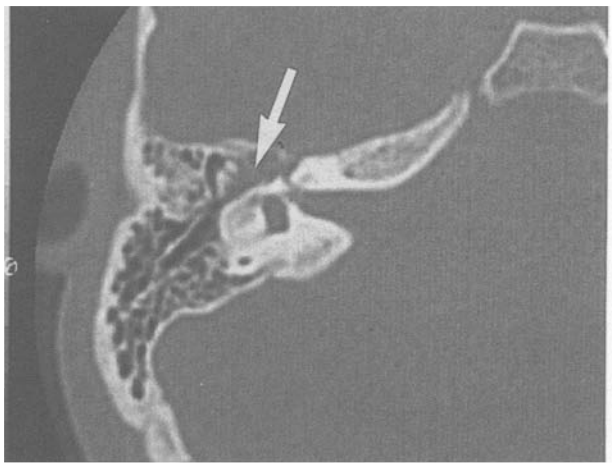

FIG. 1
CT scan showing tumour (arrow) in the right epitympanum lying medial to the body of the incus and malleus head.

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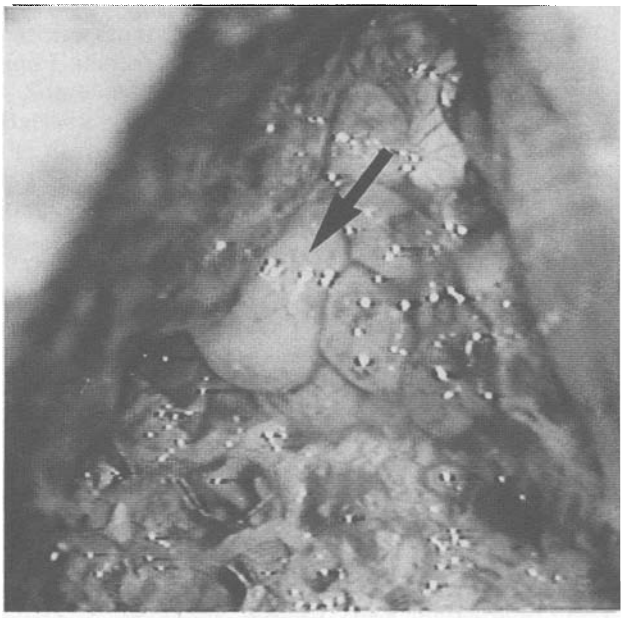

FIG. 2
Video photographs at operation showing the tumour (arrow) exposed in the antrum via the cortical mastoidectomy.

The mastoid and middle ear were explored using a combined approach and opening the mastoid revealed a pale lobulated tumour extending into the antrum (Figure 2). The tumour was wrapped around the ossicles, including the long process of the incus and appeared to originate from the sheath of the tympanic segment of the facial nerve. On frozen section histology the tumour appeared to be a malignant small cell tumour but, in view of the uncertainty of the histology, it was decided not to sacrifice the nerve. The tumour was removed and complete macroscopic clearance was achieved, leaving the facial nerve and ossicular chain intact. Post-operatively, there was early evidence of facial nerve recovery, with complete recovery in two months. Histology showed the tumour to be a neuroblastoma arising from the facial nerve. No tumour was found elsewhere and the patient received a post-operative course of radical radiotherapy to the right temporal bone. He required a ventilation tube in the right tympanic membrane for a middle ear effusion, but retained good hearing. At review 39 months later he remained well and free from tumour.

Pathology

Paraffin sections showed a malignant tumour composed of small undifferentiated cells with dark nuclei and negligible cytoplasm (Figure 3). The differential diagnosis, therefore, included primitive neuroectodermal tumour (PNET) and related lesions, rhabdomyosarcoma and Ewing's sarcoma. Detailed immunocytochemical staining gave positive reactions with neuronspecific enolase (NSE) and vimentin: there was negative reaction to a wide range of other markers (Table I). These findings are suggestive of a neural tumour. Small cell neural tumours are classified as PNET if they are of central origin and neuroblastoma if they are of peripheral origin. Since this tumour arose from a peripheral nerve and lacks astrocytic markers it has been classified as a neuroblastoma.

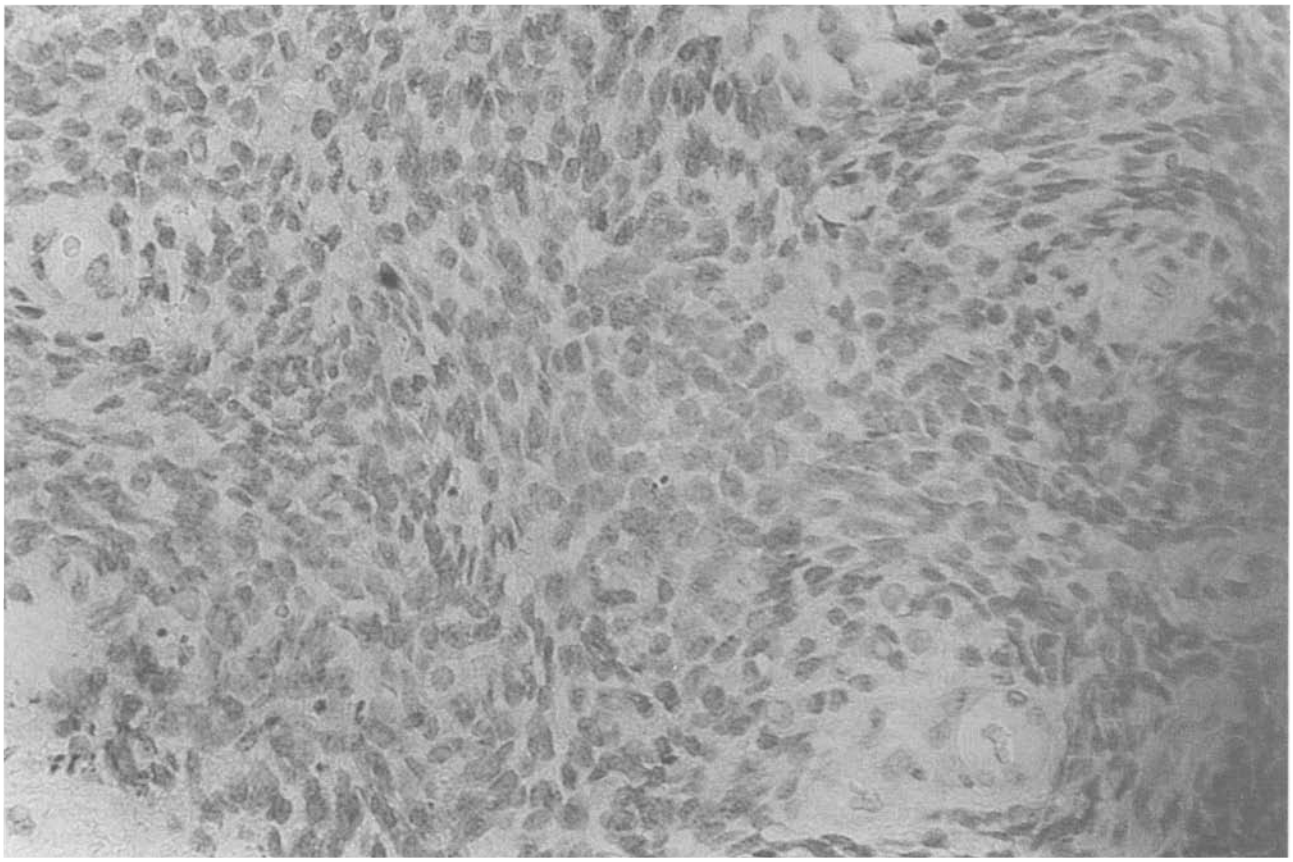

FIG. 3
Histological appearance of the tumour, showing small round cells. (H&E; × 300).

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TABLE I
REACTIONS TO IMMUNOCYTOCHEMICAL STAINS

Antibody	Detects	Staining
ASMA	α-smooth muscle actin	–ve
CAM 5.2	Low molecular weight cytokeratins	–ve
Chromogranin	Neurosecretory granules	–ve
Desmin	Intermediate filament of muscle cells	–ve
GFAP	Glial fibrillary acidic protein	–ve
LCA (CD45)	Leucocyte common antigen	–ve
MB2	B-cell marker	–ve
NSE	Neurone specific enolase	+ve
PGP 9.5	Neural-associated marker	–ve
S100	Neural-associated marker	–ve
Synaptophysin	Neural marker	–ve
Vimentin	Intermediate filament of mesenchymal cells	+ve

Discussion

Neoplasia is an uncommon cause of facial paralysis and it is estimated that it represents about five per cent of all cases (Shambaugh and May, 1980). It may result from primary facial nerve tumours, involvement from other temporal bone neoplasms or metastases from a distant primary. Of approximately 150 primary facial nerve tumours reported to date, the great majority are histologically benign (Sneige and Batsakis, 1991). In children, facial nerve paralysis is predominantly the result of infection, trauma (at birth or later) and congenital disorders. Paralysis of neoplastic origin is less common, and in one paediatric oncological hospital of over 350 beds only 22 cases of facial paralysis of malignant aetiology were noted over a 10-year period (Jung and Gutjahr, 1976).

A primary tumour of the nerve is well down the list in the differential diagnosis of a patient presenting with a facial paralysis – in one series primary tumours accounted for only five (0.7 per cent) of 552 cases (Kobayashi, 1979). Approximately 40 per cent of all facial paralyses are idiopathic (May and Hardin, 1977), but not until the other causes of lower motor neurone facial paralysis have been ruled out, can a diagnosis of Bell's palsy be made. By definition, this is a diagnosis by exclusion.

The diagnosis of a tumour in a patient with a facial nerve paralysis depends on a thorough neuro-otological investigation, coupled with a high index of suspicion. There is a wide variation in the presenting signs and symptoms. Paralysis of the nerve occurs in almost 50 per cent of patients with a primary tumour of the nerve, and the paralysis is more frequent when the tumour is intratemporal – occurring in 84 per cent of patients in one series (Janecka and Conley, 1987). The paralysis is classically of gradual onset and slowly progressive. May and Hardin (1977) have shown that maximal degeneration of the nerve in Bell's palsy occurs within two weeks, and a unilateral paralysis progressing beyond three weeks strongly suggests a neoplastic aetiology. Sudden onset paralysis, however, does not rule out tumour aetiology, and in one series 20 per cent of tumours presented as paralysis of sudden onset (Fisch and Ruttner, 1977). May and Hardin (1977), in a series of 20 patients with ipsilateral recurrent facial paralysis, noted that six (30 per cent) were of neoplastic origin. All six of these had been initially misdiagnosed as Bell's palsy. Recurrence of facial paralysis ipsilaterally should therefore be considered as suggestive of tumour aetiology. A number of other clinical features should alert the clinician to the possibility of a neoplastic aetiology (Jackson *et al.*, 1980). These are: slow progression beyond three weeks, no return of function after six months, facial hyperkinesia or spasm, other cranial nerve involvement, single branch paralysis and pain.

High-resolution CT or MRI scanning are the diagnostic investigation of choice, giving a detailed evaluation of the temporal bone and facial nerve, and should be used in any cases showing the above features.

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