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Intraparotid facial nerve schwannoma in a child

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Abstract
Neoplasms of the facial nerve presenting as a parotid mass are uncommon. In the absence of a facial palsy their origin from the nerve is usually diagnosed intraoperatively. The majority of these neurogenic neoplasms are schwannomas, with neurofibromas occurring rarely. Although the Schwann cell is the key element in both, they have distinct histopathological characteristics, and their clinical course and management often differs. The first reported case of an intraparotid facial nerve schwannoma in a child in the English literature is presented.

Key words: Parotid gland; Facial nerve; Schwannoma; Child

Introduction

Schwannomas (neurilemmomas) are benign tumours which arise from the sheath of peripheral and cranial nerves. Facial nerve schwannomas can arise from any segment of the nerve, from the glial-Schwann cell junction at the cerebellopontine angle to the peripheral branches in the face. Rarely do they arise from the intraparotid portion of the facial nerve (Jung *et al.*, 1986). We present such a case in an eight-year-old girl, who underwent surgical excision of the tumour with preservation of facial nerve function, and review the literature.

Case report

An eight-year-old girl presented to the ENT Department with a six-week history of a hard lump in the left preauricular region. There was no history of facial weakness. Physical examination revealed a 2×1 cm firm, nontender, mobile mass anterior to the tragus and facial function was normal. Fine needle aspiration cytology was inconclusive and routine blood tests were normal. A superficial parotidectomy with frozen section control was planned. A cervicofacial incision was made, skin flaps were raised and a 4×3 cm mass was visible. Frozen section biopsy showed this to be a benign schwannoma. The mass was dissected, with ease, off all branches of the facial nerve except for the temporal branch, to which it was adherent. However, it was dissected free of this branch with apparent anatomical preservation of the nerve. Post-operatively there was transient weakness of the temporal branch of the facial nerve which gradually resolved and on review three months later facial function was normal.

Pathology

The resected tumour was irregularly elongated and nodular measuring 4×1.5 cm. It appeared encapsulated and, on sectioning, was found to be uniformly solid with a cream and pale grey cut surface. Microscopically it showed compact fascicles of spindle cells with prominent nuclear palisading, corresponding to Antoni A type tissue (Figure 1). Almost all of the tumour exhibited this pattern quite uniformly, there being only minute foci with the looser Antoni B structure. There was no nuclear hyperchromatism and pleomorphism and mitotic figures were not found (Figure 2). Tumour cells diffusely and strongly expressed S-100 protein, confirming the diagnosis of schwannoma.

Discussion

Tumours arising from the intraparotid portion of the facial nerve are rare (Neely and Alford, 1974; Jung *et al.*, 1986; Sullivan *et al.*, 1987; Kayem *et al.*, 1995). Benign neoplasms are of two types: schwannoma (neurilemmoma, neurolemmoma, neurinoma) and neurofibroma. Schwannomas are different from neurofibromas in terms of their histological appearance and biological behaviour. The schwannoma is a solitary, encapsulated tumour, usually attached to a nerve, that appears to push axons aside as it

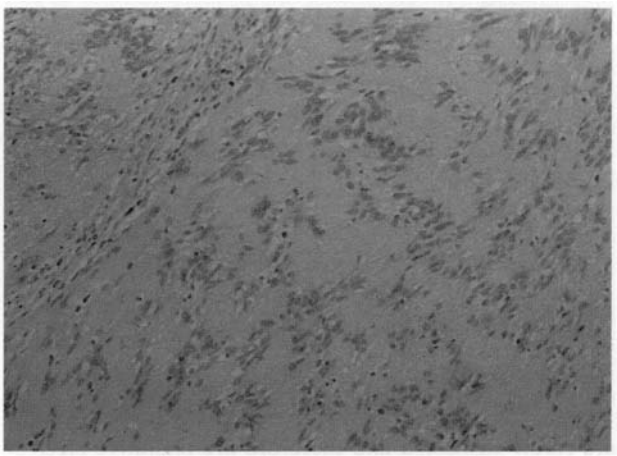

FIG. 1
Facial nerve schwannoma. Note the prominent nuclear palisading. (H & E; × 100)

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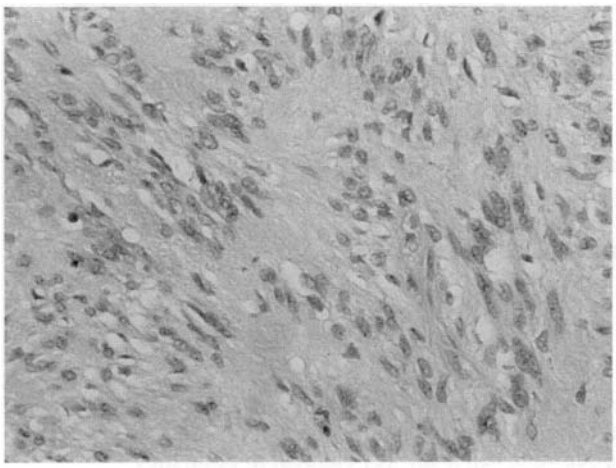

FIG. 2
Nuclei are uniform and mitoses are absent (H & E; × 250)

grows and therefore surgical removal with preservation of the nerve is usually possible. Microscopically two patterns may be seen: a compact spindle-cell pattern with nuclear palisading forming Verocay bodies (Antoni type A) admixed in variable proportions with a looser and often more pleomorphic spindle-cell tissue with foam cells (Antoni type B) (Batsakis, 1979; Sullivan *et al.*, 1987).

Neurofibromas on the other hand, are nonencapsulated, may be diffuse or plexiform, and are composed of an admixture of all elements of a peripheral nerve i.e. axons, Schwann cells, fibroblasts and (in the plexiform type) perineural cells. They do not contain Verocay bodies (Batsakis, 1979). These lesions often intimately involve the nerve, making surgical resection with preservation of the nerve of origin difficult. Malignant degeneration rarely occurs in schwannomas but is a relatively common feature of neurofibromas (Kayem *et al.*, 1995). Neurogenic sarcomas have been found in five to 15 per cent of patients with type 1 neurofibromatosis (Sullivan *et al.*, 1987; Prasad *et al.*, 1993). There has been no recorded instance of sarcomatous degeneration of a neurofibroma involving the facial nerve, but malignant schwannomas involving this site have been reported (Conley and Janecka, 1974; Sullivan *et al.*, 1987), including one in a child (Hasan and Kazi, 1986).

Neurogenic intraparotid facial nerve neoplasms always present as a preauricular or facial mass (Prasad *et al.*, 1993). Nearly half of these extratemporal tumours involve the main trunk of the nerve (Sullivan *et al.*, 1987). Hemifacial spasm or twitching suggests the presence of a compressive or infiltrative lesion and may precede facial paralysis by weeks or months (Neely and Alford, 1974; Pillsbury *et al.*, 1983; Wiet *et al.*, 1983). Facial paralysis associated with a parotid mass nearly always signals a malignancy (Jackson *et al.*, 1980) but facial paresis resulting from an extratemporal facial nerve schwannoma has been reported (Conley and Janecka, 1974; Neely and Alford, 1974; Prasad *et al* 1993). For neurogenic tumours involving the peripheral portion of the facial nerve system, intra-operative visual inspection of the main trunk of the facial nerve and all its peripheral branches has been recommended, to rule out multicentricity (Conley and Janecka, 1974). Neurogenic neoplasms in the parotid gland are rarely suspected and therefore pre-operative electrodiagnostic tests are often not considered. Electroneuronography may show subclinical neoplastic involvement of the facial nerve (Neely and Alford, 1974).

The histological difference between schwannomas and neurofibromas of the facial nerve may dictate the type of surgical resection which is possible. Theoretically, schwannomas can be stripped free of the nerve without sacrificing it, as in this case. In contrast, fibres from the facial nerve pass directly through a neurofibroma and separation of the nerve fibres from the tumour is rarely possible (Sullivan *et al.*, 1987; Kayem *et al.*, 1995). Neurofibromas generally require nerve sectioning and cable grafting, using the sural nerve for a long defect and the greater auricular nerve for a short defect (Pillsbury *et al.*, 1983; Sullivan *et al.*, 1987).

In conclusion, while neurogenic tumours of the facial nerve in the parotid gland are rarely anticipated and electroneuronography is as yet inconclusive, they should be suspected intra-operatively if the tumour is inseparable from the nerve (Wiet *et al.*, 1983) and when electrical stimulation of the tumour elicits facial movement (Neely and Alford, 1974). Furthermore, intra-operative confirmation of the tumour by frozen section biopsy is imperative because nerve preservation can be attempted in benign tumours whereas the nerve may have to be sacrificed in malignant tumours (Neely and Alford, 1974; Pillsbury *et al.*, 1983).

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