The Journal of Laryngology and Otology
November 1992, Vol. **106,** pp. 1011–1015

View from Beneath: Pathology in Focus

Benign nasal schwannoma

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Abstract
Nerve sheath tumours of the head and neck region mainly involve the eighth cranial nerve with only 4 per cent occurring in the paranasal sinuses. Only 32 cases of benign schwannomas occurring in the paranasal sinuses have been reported. We present a further case, review the current literature, and discuss the clinical details and specific histological features.

Case report

A 46-year-old man presented with a six month history of left sided nasal obstruction and epiphora. Recently he had developed frontal headaches. ENT examination revealed a large pale mass in the left nasal cavity. Physical examination and routine blood tests were otherwise normal. Plain X-rays of the paranasal sinuses (Fig. 1) showed opacification of the left frontal sinus, a fluid level in the left maxillary antrum and a soft tissue mass in the left nasal cavity. Computerized tomography (Figs. 2 & 3) revealed a soft tissue mass of 5 mm diameter in the left nasal cavity with deviation of the nasal septum to the right and bulging of the medial wall of the left antrum.

At examination under general anaesthesia, a pale, greyish tumour was identified coming from the region of the middle turbinate. A histological diagnosis of a benign schwannoma was made. Lateral rhinotomy was performed, revealing a large tumour in the left nasal cavity. Extension had occurred into the ethmoid sinus with erosion of the lamina papyracae and medial orbital rim. After complete removal of the tumour, the frontal sinus was drained of pus and the remaining sinuses on that side were inspected. The cavity was packed with BIPP for 48 hours and the patient was discharged home after 10 days.

The gross specimen was a tan coloured, nodular and partially encapsulated mass $4 \times 2.3 \times 1.7$ cm (Fig. 4). On sectioning it was haemorrhagic with myxoid areas. Histologically the tissue consisted of areas showing a fibrillary appearance, with focal verocay bodies (Fig. 5). In addition, there were other areas showing chronic inflammation and fibrosis. The S 100 protein immunostaining was strongly positive both in the nuclei and cytoplasm of the tumour cells (Fig. 6). On electron microscopy, there was a dense basal lamina, and redundant stacks of collagen between the tumour cells (Fig. 7).

Discussion

Nerve sheath tumours occur in the head and neck region more frequently than any other part of the body, mainly affecting the eighth cranial nerve (Dasgupta *et al.*, 1969; Batsakis, 1979; Wilson *et al.* 1988). However, only 4 per cent involve the paranasal sinuses (Shugar *et al.*, 1981). Benign schwannomas are the commonest type, with only 32 cases occuring within the paransal sinuses having been reported (Iwamura *et al.*, 1972; Robitaille *et al.*, 1975; Shugar *et al.*, 1981; Mahe *et al.*, 1983; Ross *et al.*, 1988; Kautzky and Schenk, 1989; Hilstrom *et al.*, 1990).

Benign schwannomas originate from the nerve sheath which is composed of schwann cells and fibroblasts. Harkin and Reed

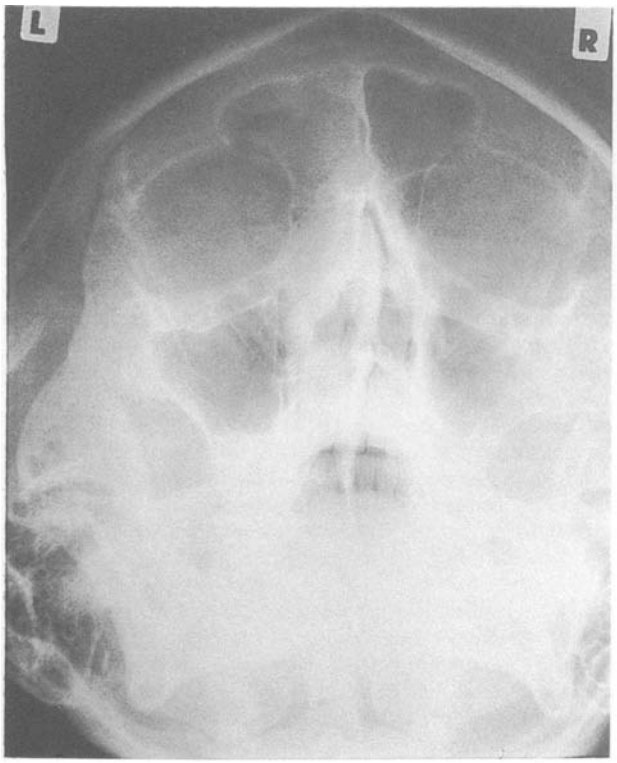

FIG. 1
Plain X-rays of the paranasal sinuses.

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Accepted for publication: 29 June 1992.

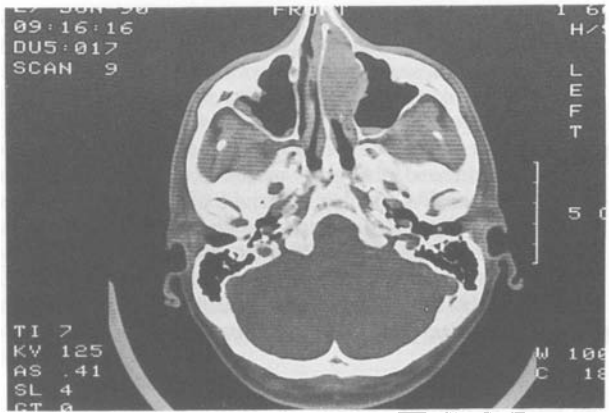

FIG. 2

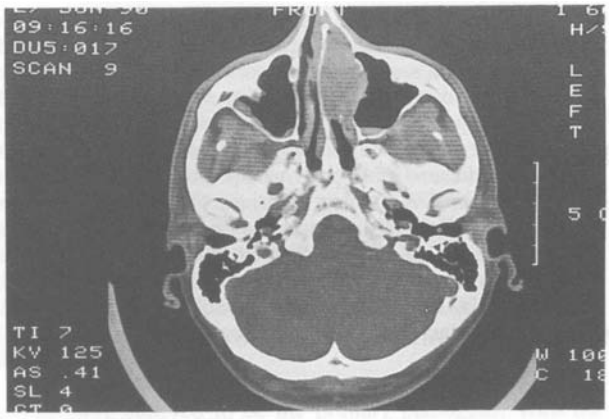

FIG. 3

FIGS. 2 and 3
CT scans of the nose and paranasal sinuses.

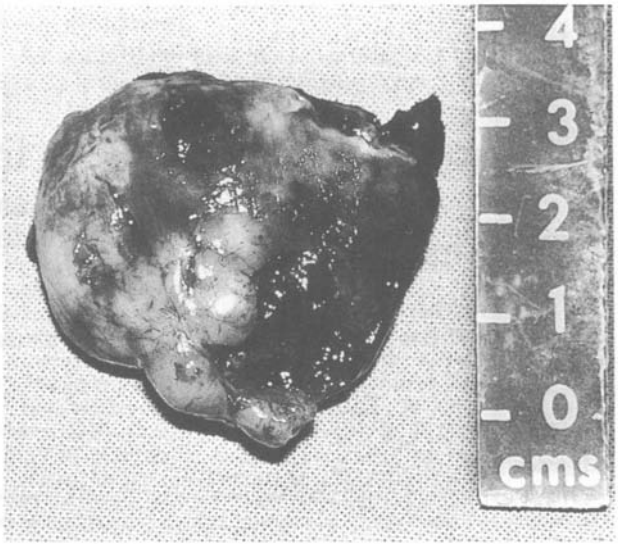

FIG. 4
Surgical specimen.

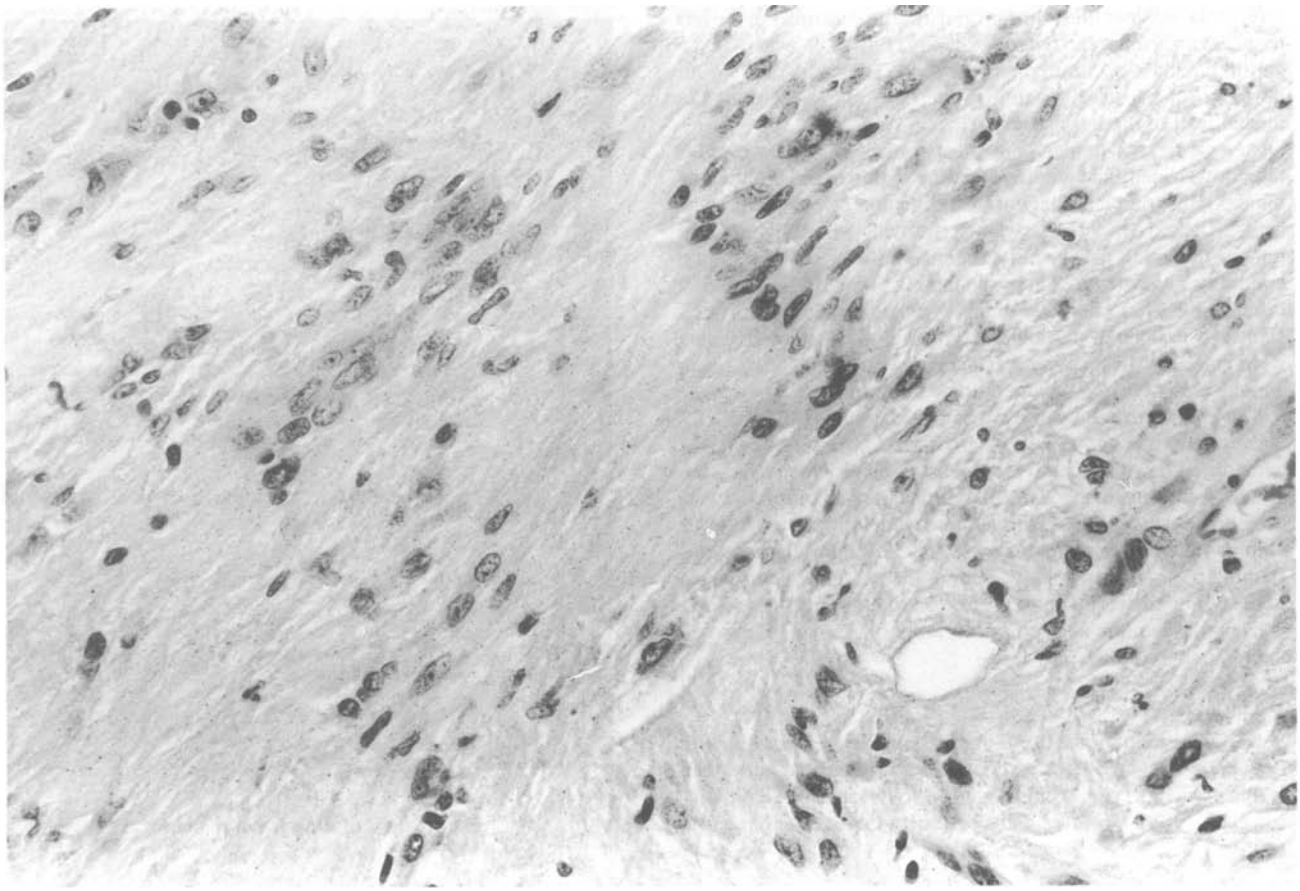

FIG. 5
Verocay body with aligned nuclei.

https://doi.org/10.1017/S0022215100121644 Published online by Cambridge University Press

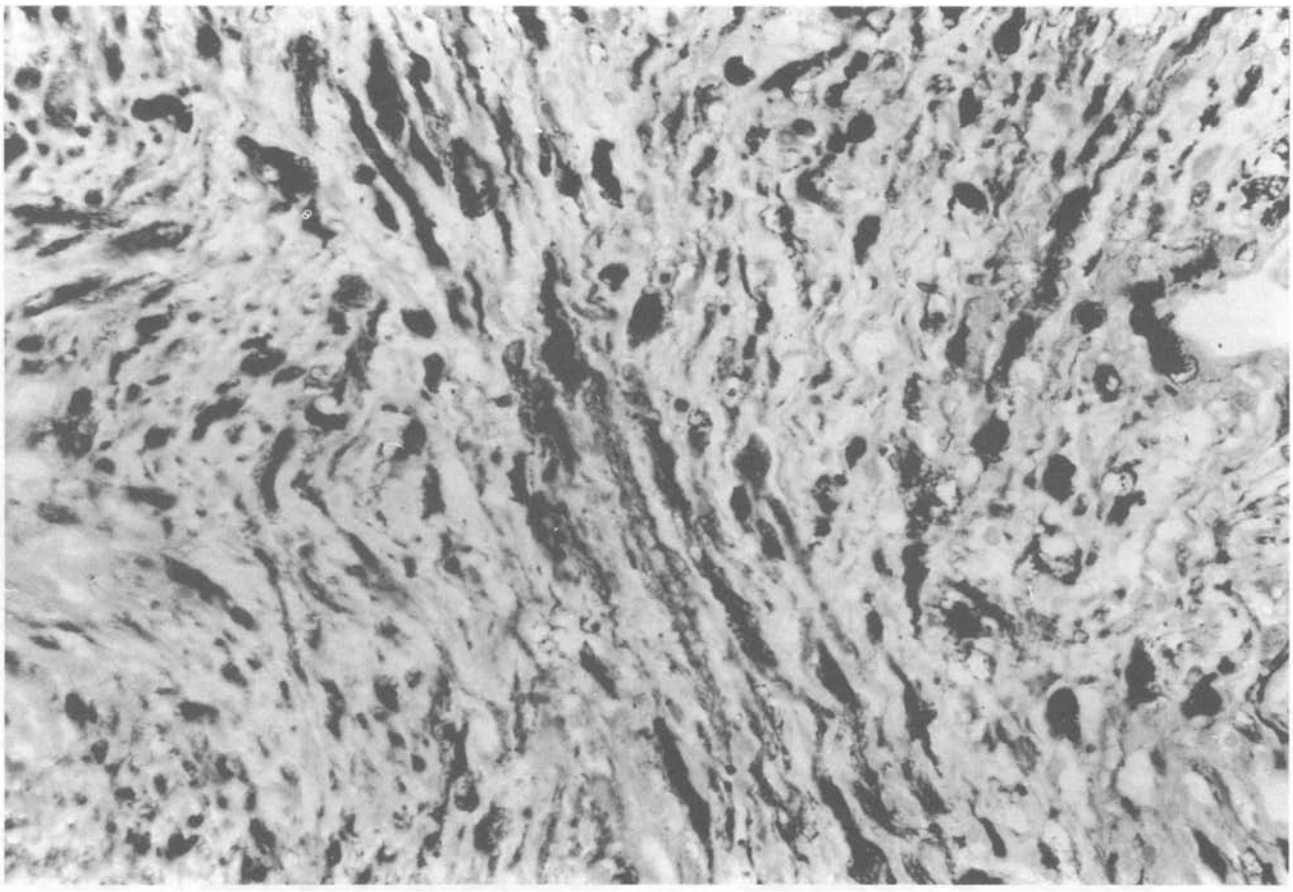

FIG. 6
S 100 protein positive immunohistochemical staining.

(1969) and Batsakis (1979) have reported that as both these cell types are derived from neuroectoderm, either could give rise to the tumour. This has probably contributed to the confusing array of synonyms that have been previously used, including neurilemomas, neurilemmomas, neurolemomas and neuromas. The schwann cell is now accepted as the most likely source of all nerve sheath tumours (Dasgupta *et al.*, 1960; Batsakis, 1979). Closely related to schwannomas are neurofibromas and although Perzin *et al.* (1982) stated that they may be distinguishable histologically, Horak *et al.* (1983) found that histological features of both tumours may be frequently seen in the same surgical specimen. Therefore electron microscopy and immunohistochemical analysis are often necessary to diagnose and accurately classify nerve sheath tumours (Hillstrom *et al.*, 1990). It was felt important to differentiate between tumour types as malignant change may occur in neurofibromas but not schwannomas (Harkin and Reed, 1969; Perzin *et al.*, 1982; Bruner, 1987). Recently, however, Rasbridge *et al.* (1989) have reported malignant change in a long-standing benign schwannoma.

Apart from the olfactory nerve, any of the nerves supplying the nose and paranasal sinuses may give rise to the tumour, somatic or autonomic. It is, however, unusual to identify the particular nerve of origin. The sites of origin are nasal-ethmoid, maxillary sinus, intranasal cavity and sphenoid sinus in order of frequency (Batsakis, 1979).

Clinical presentation is not confined to particular age, sex or racial groupings (Calcaterra *et al.*, 1974). The tumour is slow growing, tending to expand and erode surrounding bony cavities. Therefore symptoms and signs depend upon the site of origin and subsequent involvement of surrounding structures. More common non-specific symptoms include nasal airway obstruction, hyposmia, rhinorrhoea, exopthalmos and facial swelling. Epistaxis is associated with nasal-ethmoid tumours and pain with maxillary sinus lesions (Robitaille *et al.*, 1975). Skull base involvement by sphenoid sinus tumours can produce cranial nerve deficits and facial pain. Posterior extension into the pituitary fossa may result in hypopituitarism (Calcaterra *et al.*, 1974). Involvement of the lacrimal apparatus resulting in epiphora has not been previously described but given the nature of the tumour, this symptom is not unexpected.

A variety of imaging techniques can be used to demonstrate and demarcate schwannomas. Plain X-ray films of the region can demonstrate evidence of bone thinning or erosion, a soft tissue mass or sinus obstruction. Computerized tomography is the examination of choice and should include the intracranial cavity as intracranial extension has been reported (Zovickian *et al.*, 1986). The use of magnetic resonance imaging (MRI) in this condition has not yet been fully evaluated. In a recent review, Lloyd (1989) states that MRI scanning of the paranasal sinuses is very useful in differentiating tumour from retained secretions. However, a major disadvantage of this technique is the poor demonstration of bone and because of this high resolution CT scanning is recommended.

Schwannomas histologically have two appearances: Those with a cellular component (Antoni A) and those with a loose myxoid component (Antoni B). Alternating Antoni A and B areas are seen in varying combinations. The Antoni A areas are composed of compact spindle cells with twisted nuclei, indistinct cytoplasmic borders, and occasional intranuclear vacuoles. The cells are arranged in short bundles, or interlacing fascicles. If highly differentiated they may show nuclear palisading, whorling of cells, and verocay bodies (compact groups of parallel spindled-shaped nuclei—Fig. 5). The Antoni B areas are far less orderly and cellular. Glands and benign epithelial structures have been reported rarely (Fletcher *et al.*, 1986). About 5 per cent of schwannomas grow in a plexiform or multi-nodular pat-

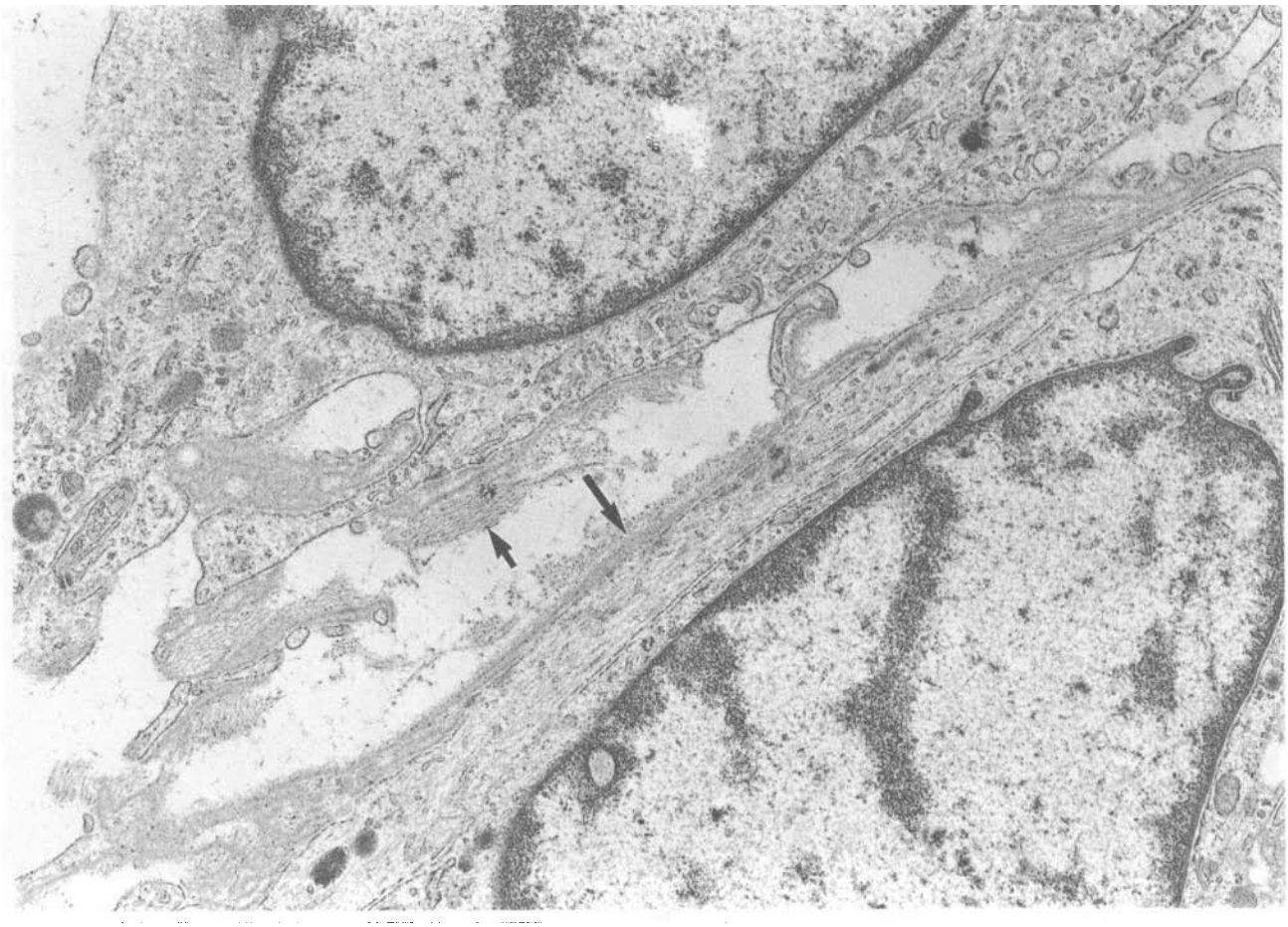

FIG. 7

Electron micrograph showing electron dense basal lamina coating the surface of schwann cells (large arrow), and lying in redundant stacks (small arrow).

tern, which may or may not be apparent macroscopically (Woodruff *et al.*, 1983; Fletcher and Davies, 1986).

Ultrastructural analysis has shown that unlike neurofibromas which have a mixture of cell types, schwannomas consist almost exclusively of schwann cells (Erlandson and Woodruff, 1982). It would appear from the ultrastructural features that Antoni B areas are degenerating Antoni A areas (Lyser, 1975; Lazarus and Trombetta, 1978; Kautzky and Schenk, 1989). Using immunohistochemical techniques it is possible to identify cell types not recognizable by morphological features. S 100 protein (S 100) is a neural-crest marker antigen present in the supporting cells of the nervous system (Nakajima *et al.*, 1982; Steffanson *et al.*, 1982). It is an important diagnostic tool as schwannomas show intense immuno-staining for S 100 (particularly Antoni A areas). Neurofibromas stain less intensely and other connective tissue tumours not at all (Weiss *et al.*, 1983).

The appropriate treatment of the tumour is complete surgical removal (Shugar *et al.*, 1982). To some extent the surgical approach is determined by the tumour site. For most cases a lateral rhinotomy affords the best access to the nasal cavity and paranasal sinuses. Alternative procedures include a Caldwell Luc approach for maxillary sinus lesions with the addition of external ethmoidectomy if involvement of the ethmoidal system has occurred. Skull base surgery may be necessary for sphenoidal schwannomas. More advanced lesions or any tumour extending through the cribiform plate are more safely tackled via a craniotomy (Calcaterra *et al.*, 1974), combined with any of the other approaches.

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Key words: Paranasal sinus neoplasms, benign nasal schwannoma