The Journal of Laryngology and Otology
February 1997, Vol. **111**, pp. 145–147

Diffuse neurofibroma obstructing the external auditory meatus

DARRAGH COAKLEY, F.R.C.S.I., MARCUS D. ATLAS, F.R.A.C.S.

Abstract
A case is presented of a 36-year-old male with narrowing of the external meatus due to a diffuse neurofibroma. This unusual variety of neurofibroma spreads superficially and has many ectatic blood vessels. The size, vascularity, uncertain edges and a tendency to recur makes surgical removal difficult. The treatment options are discussed.

Key words: Ear canal; Neurofibroma

Introduction

Neurofibroma is a relatively common tumour and is classified as solitary or multiple. Solitary lesions are not usually associated with systemic manifestations unlike multiple lesions which are often seen in patients with neurofibromatosis or von Recklinghausen's disease (Requene and Sanqueze, 1995). There are a number of histological subtypes of neurofibroma including localized, plexiform and diffuse. The diffuse variety is uncommon and little has been reported on this variant of neurofibroma (Megehed, 1994).

Case report

A 35-year-old male presented with a long history of left-sided wax impaction and secondary otitis externa. Examination revealed a soft tissue narrowing of the left cartilaginous external meatus. He had no history of trauma but appeared to have a diffuse periauricular swelling. The audiogram (Figure 1) showed a 40 dB conductive loss in the left ear with headphones but normal hearing with an insert. A computed tomography (CT) scan (Figure 2) showed a mass of density similar to muscle encroaching on the external canal from the posterior aspect. This mass extended up over the lateral aspect of the squamous temporal bone and was associated with shallow erosion of the lateral aspect of the mastoid cortex. Surgery was recommended to biopsy the lesion and perform a meatoplasty if possible.

A post-auricular incision was made and it was immediately apparent that there was a pale amorphous mass of soft tissue over the mastoid process and extending up to the inferior temporal bone. It was adherent to the bony cortex and to the skin of the meatus and ear canal. Marked bleeding occurred and a vascular tumour was suspected. Frozen sections were obtained and the preliminary report suggested the possibility of a haemangioma of muscle. Complete excision of the lesion would have involved removal of the pinna, canal and periauricular skin up to the inferior temporal line. The lesion was partially excised but the meatal skin was not excised to avoid the possibility of seeding of tumour.

Histopathology

Paraffin sections showed the tumour to be a neurofibroma of the diffuse type (Figure 3). The tumour cells had ovoid to elongated, often wavy nuclei randomly distributed in a matrix of fine undulating collagen fibres. Clusters of Meissner-like bodies were present throughout; these were mistaken, in the frozen section, for skeletal muscle fibres. Nests of adipose tissue, probably pre-existing were surrounded and infiltrated by tumour. Immunoperoxidase stains for S100 protein showed positive staining of tumour cell nuclei and cytoplasm, but stains for neurofilaments and myelin basic protein showed only occasional nerve twigs within the tumour, reflecting the growth of diffuse neurofibromas beyond the perineurium of the nerve of origin. The tumour was extremely vascular. There were

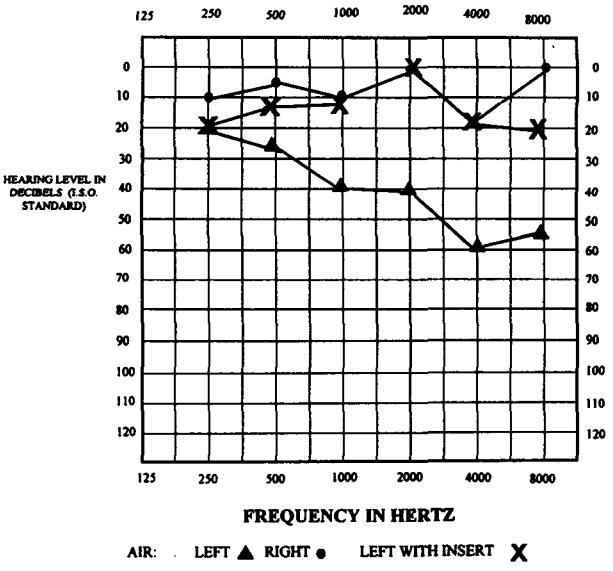
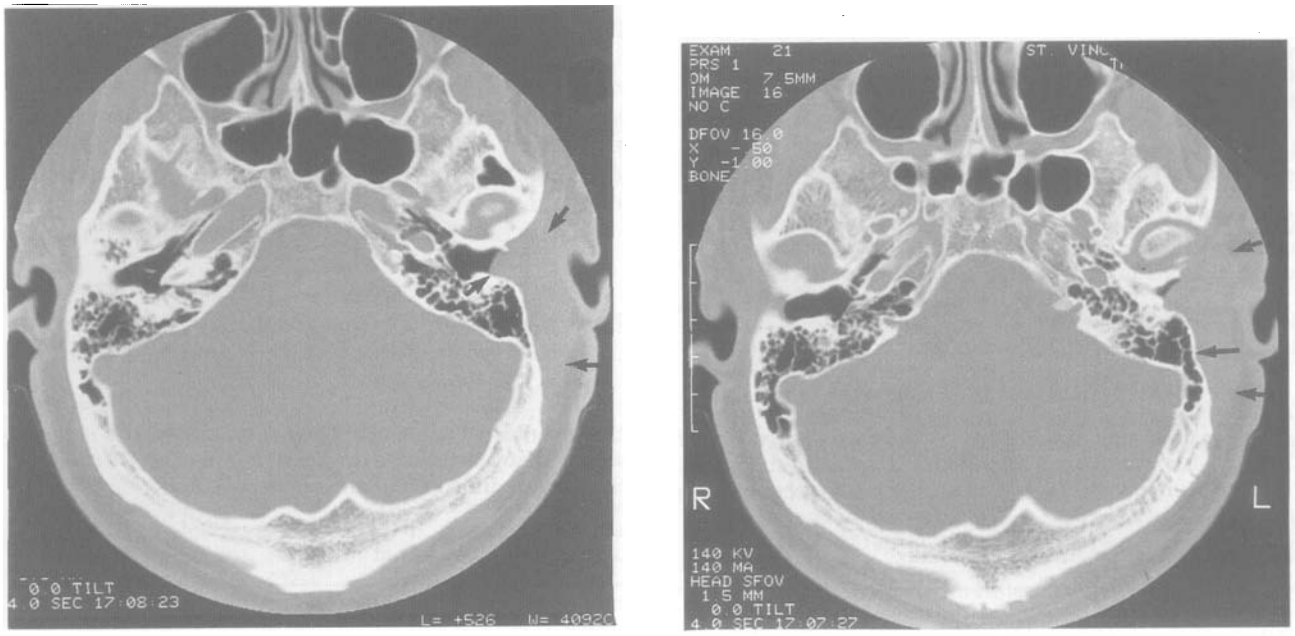


FIG. 1
Air conduction audiogram showing change of hearing with insert in left ear.

From the Department of Ear and Skull Base Surgery, St. Vincent's Hospital, Sydney, Australia.
Accepted for publication: 16 November 1996.



(a) (b)

FIG. 2

Soft tissue mass overlying mastoid cortex and squamous temporal bone causing shallow erosion of cortex and narrowing of cartilaginous external canal.

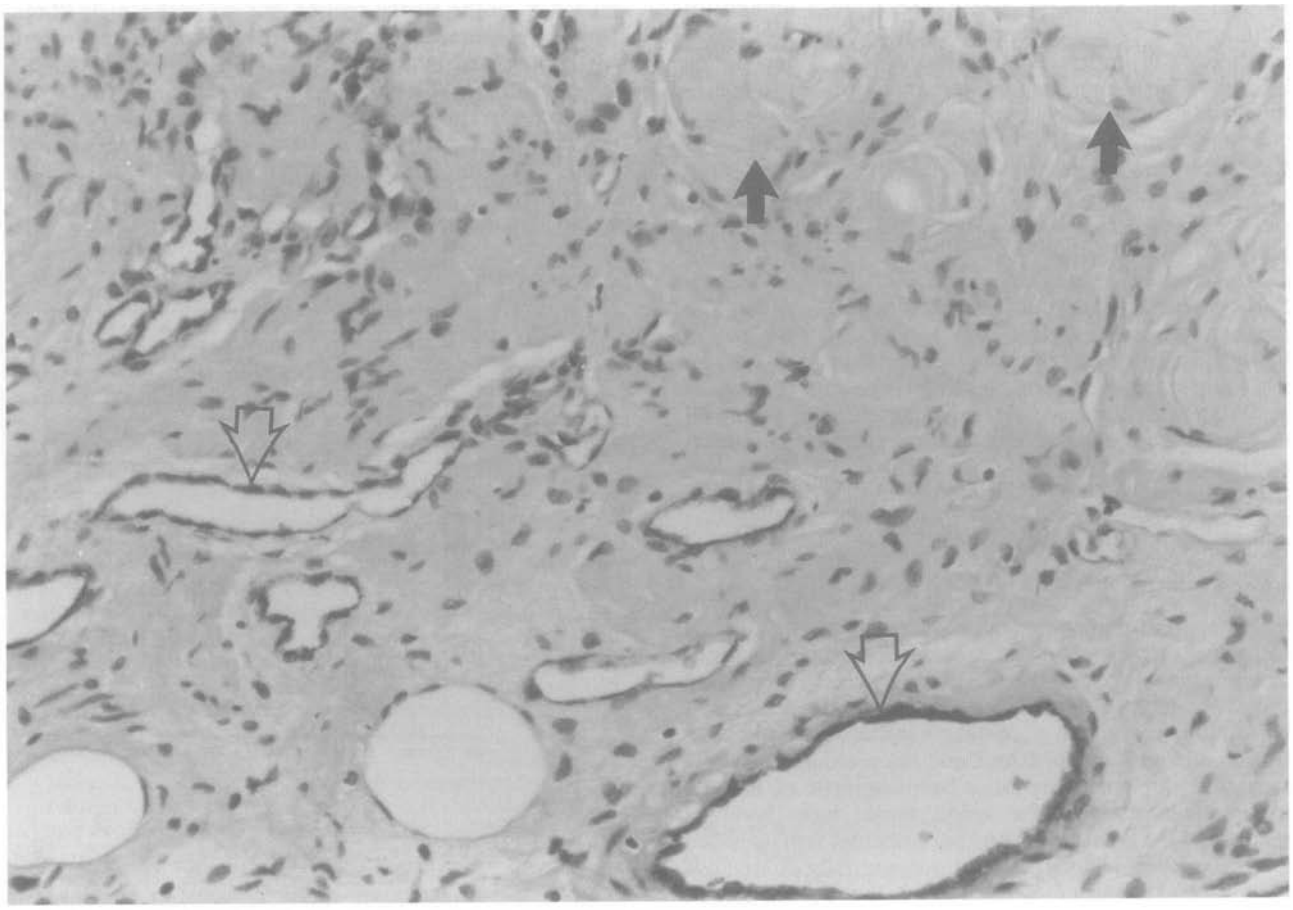


FIG. 3

Photomicrograph of the tumour showing Meissner-like corpuscles (black arrows) and staining of the endothelial cells of the prominent vessels (clear vessels) with an antibody to Factor VIII related antigen (Immunoperoxidose stain, original magnification × 25).

https://doi.org/10.1017/S0022215100136680 Published online by Cambridge University Press

large, ectatic, irregular vascular spaces, sometimes with a few muscle fibres in their walls, as well as small capillary blood vessels.

Comments

Diffuse neurofibroma is an uncommon variant of neurofibroma that occurs primarily in young adults and children. A mean age of 60 years has been reported in one series in which the periauricular area was the most commonly involved site in the head and neck region (Megehed, 1994). Between 10 per cent (Enzinger and Weiss, 1993) and 60 per cent (Megehed, 1994) of these tumours are associated with neurobroma type I. Malignant change is rare (Enzinger and Weiss, 1993). Extension beyond the perineurium is usual and some authors have called these lesions paraneurofibroma to highlight this (Harkin and Reed, 1968). The tumour is ill-defined and greyish, spreading extensively through the connective tissue septae and between fat cells, enveloping rather than destroying structures. In some of these tumours mature fat or large ectatic blood vessels are mixed with the neurofibromatous element in a complex arrangement (Enzinger and Weiss, 1993). The large ectatic blood vessels may be the dominant feature. The formation of aggregates of tumour cells into tactoid bodies or Meissner bodies (Taxby, 1990) is a characteristic feature and with S100-positivity distinguishes this tumour from dermatofibrosarcoma protuberans. Ultrastructurally these lesions have been shown to be composed of Schwann cells (Theaker and Fletcher, 1989).

This patient does not have any other evidence of NF1. Excision of the lesion is not mandatory as malignant change is rare. Complete excision in this case would require extensive resection of muscle, skin, pinna and external auditory canal followed by reconstruction. These tumours frequently recur and treatment is usually palliative (Trevisiani *et al.*, 1982). In this case there is no significant cosmetic defect and his symptoms can be partially and simply alleviated at present.

He is to be followed up at regular intervals in the clinic with radiological evaluation and radical excision will be carried out if there is evidence of tumour growth.

Acknowledgment

The authors thank Dr J. Turner, anatomical pathologist, for her assistance.

References

Requene, L., Sanqueze, O. P. (1995) Benign neoplasms with neural differentiation: A review. *American Journal of Dermatopathology* **17(1):** 75–96.

Megehed, M. (1994) Histopathological variations of neurofibroma. A study of 114 lesions. *American Journal of Dermatopathology* **16(5):** 486–495.

Enzinger, F. M., Weiss, S. W. (1993) Ch. 24: Benign tumours of peripheral nerves. In: *Soft Tissue Tumours.* The CV Mosby Company, St. Louis, Toronto, London, pp. 610–612.

Harkin, J. C., Reed, R. J. (1968) Tumours of the peripheral nervous system (supplement). Ed. Firminger Herkes. *Atlas of Tumour.* Pathology 2nd series. Fascicle 3. Armed Forces of Institute of Pathology. S24.

Taxby, J. B. (1990) Tumours of neural differentiation. In: *Pathology of the Skin.* Chap. 51. (Farmer, E. R., Hoad, A. F., eds.), pp. 795–796.

Theaker, J. M., Fletcher, C. D. (1989) Epithelial membrane antigen expression by the perineural cell. Further studies of peripheral nerve lesions. *Histopathology* **14(6):** 581–592.

Trevisiani, T. P., Pohl, A. L., Matloub, H. S. (1982) Neurofibroma of the ear. Function and aesthetics. *Plastic Reconstructive Surgery* **70(2):** 217–219.

Address for correspondence:
Dr M. D. Atlas,
Level 10, St. Vincent's Clinic,
438 Victoria Street,
Darlinghurst NSW 2010,
Australia.