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Nasal Paraganglioma

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Introduction

Paraganglioma of the head and neck most commonly presents as either a carotid body or a jugulotympanic tumour but can also present in other sites. A case of nasal paraganglioma is reported and the treatment of this and previous cases is discussed.

Case Report

A 56-year-old lady was first seen in 1965 complaining of nasal obstruction, discharge and intermittent brisk epistaxis on the right side. Examination revealed ulceration and destruction of the right inferior turbinate. There were no other abnormalities and no signs of metastatic disease. Radiographs showed right-sided nasal occlusion but no involvement of the antrum. Biopsy confirmed a malignant neoplasm which was treated as a carcinoma with external radiotherapy to a dose of 4600cGy in fourteen fractions

In 1972 a polyp was noted on the anterior medial surface of the right inferior turbinate remnant. This polyp was biopsied and subsequently excised. The tissue was misdiagnosed as inverted papilloma. The nose healed satisfactorily after apparently complete local excision and, apart from a left-sided epistaxis from Little's area in 1975, was normal until 1976 when another polyp was seen arising from beneath the right inferior turbinate remnant. This was excised and submitted for light and electron microscopy. These confirmed a tumour of chemoreceptor origin. All available blocks from the earlier specimens were reviewed and the characteristic light microscopic appearances of paraganglioma were found in all specimens (Professor J.O'D. McGee, Oxford).

Further microscopy (Professor I. Friedmann, Northwick Park Hospital) showed vascular tumour tissue composed of fairly uniform pale-staining cells with round niclei forming strands or larger aggregations separated by fibrous septa (Figs. 1 and 2). Sections stained for reticulin showed the characteristic alveolar pattern of a paraganglioma (Fig. 3).

The nose was clear of tumour until 1982 and the patient was asymptomatic until severe epistaxis from the tumour recom-

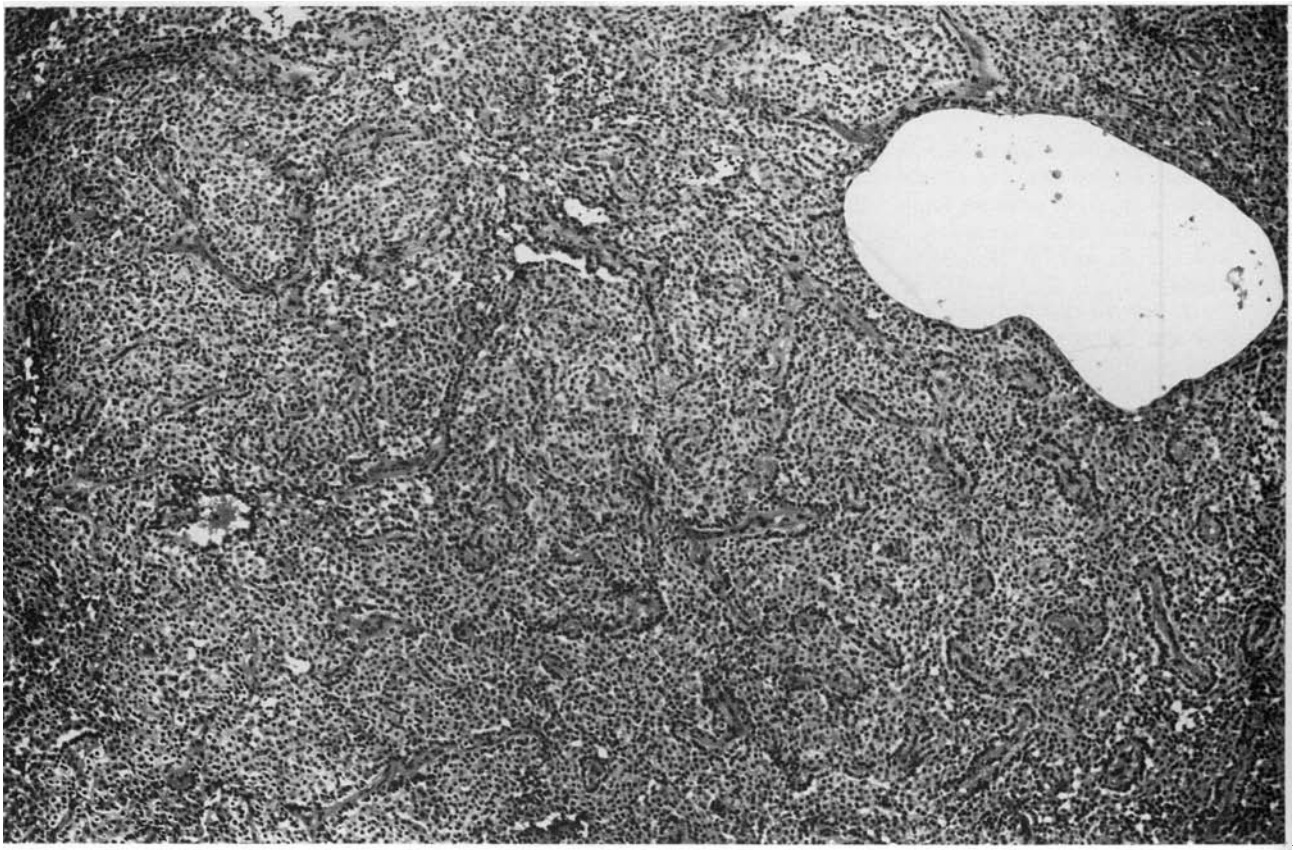


FIG. 1

Survey picture showing a solid area of the tumour composed of fairly regular round to cuboidal cells with round niclei in the pale-staining cytoplasm. The cells are closely interwoven with many capillaries. Haematoxylin-eosin x 195.

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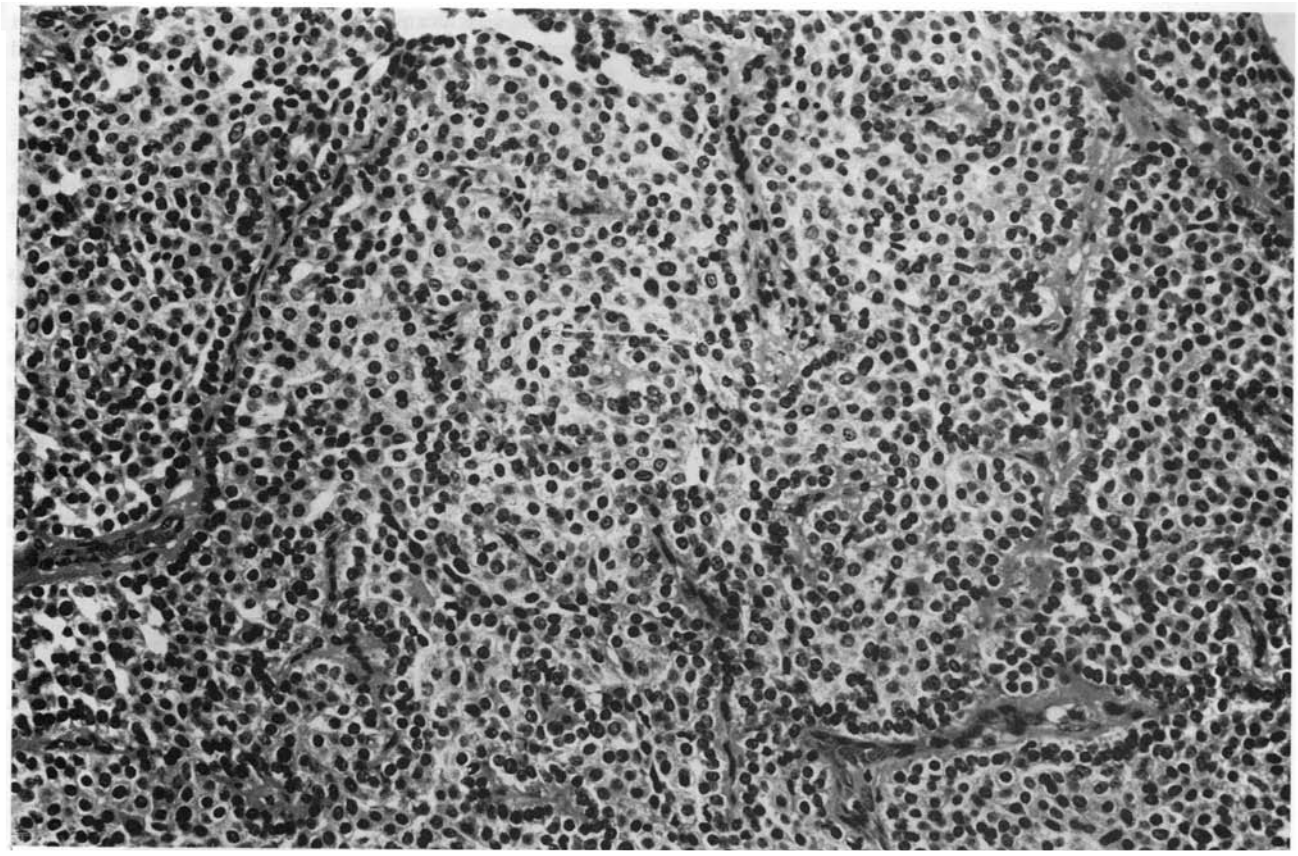


FIG. 2

Higher magnification showing the cells arranged in ribbon-like strands. Note scattered rosette-like formations in the neoplastic tissue. Haematoxylin-eosin x 400.

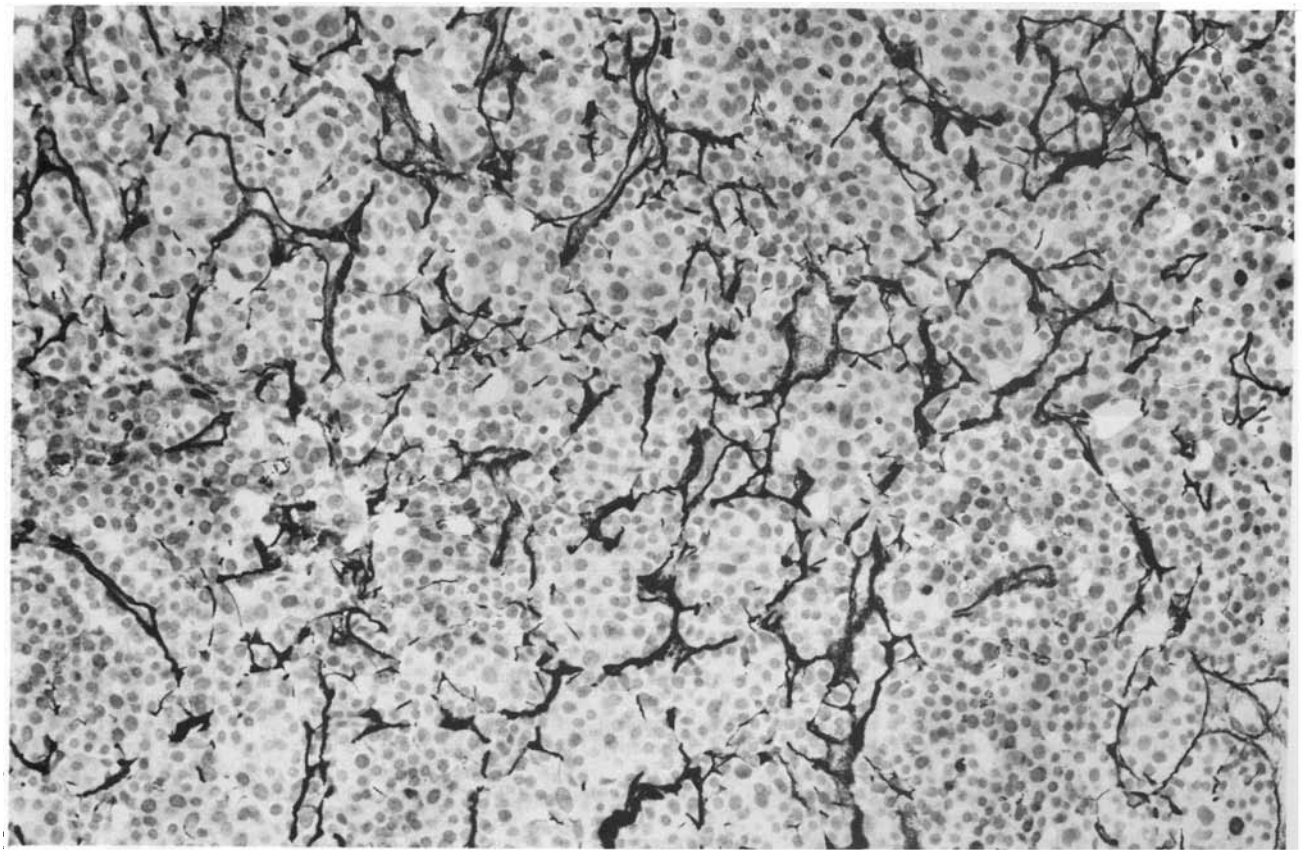


FIG. 3

Silver impregnation revealing a profuse network of reticulin and the characteristic alveolar pattern of a paraganglioma. Haematoxylin-eosin x 400.

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menced in 1984. CT scan at that time showed tumour in the nose and antrum (Fig. 4), which extended into the ethmoid air cells. The patient, by now 75 years of age, refused surgery but accepted carotid arteriography which was carried out with a view to embolisation. Tortuous carotid vessels rendered selective embolisation impossible.

After review of the original radiotherapy treatment regime, it was felt that a further low dose of radiation was appropriate. A 6cm. x 4cm. single field was therefore treated with a further 2000cGy in ten daily fractions using a Cobalt 60 machine. Treatment ended in June 1985 and, to date, there has been minimal epistaxis and no radiation necrosis.

Discussion

Paraganglioma is a tumour originating from paraganglionic cells and is divided into two different types: one is a functional chromaffinoma which produces catecholamine and the other is a non-functioning, non-chromaffin tumour. The former is most commonly seen as a phaeochromocytoma but the latter often presents in the head and neck as glomus jugulare or carotid body tumours. Other sites in the head and neck have paraganglia and may give rise to paraganglioma (Lack *et al.*, (1977).

Paraganglioma in the nose and paranasal sinuses is extremely rare and only six previous cases of definite paraganglioma arising primarily in the nose or paranasal sinuses have been reported. The presentation and features of the other cases of nasal paraganglioma are summarised in Table I.

The tumour reported by Apple and Kreines (1982) was probably a functional paraganglioma as the patient was Cushingoid on presentation and reverted to normal after irradiation of the tumour. The functional nature of the tumour is not certain, however, because the pituitary gland was also subjected to a considerable dose of radiation and it may be that this was the source of the hormonal upset. There was also radiographic evidence of residual tumour in the sinuses after treatment.

Nasal and paranasal sinus tumours seem, from the patients



FIG. 4

Axial CT scan showing tumour in the right nasal fossa and maxillary antrum.

TABLE I

REVIEW OF REPORTS OF NASAL PARAGANGLIOMA.

Patient	Site	Presenting complaint	Treatment	Outcome	Follow-up	Report
Female 89	Right anterior nares	Epistaxes	Repeated excision	Cure	2 years	Moran (1962)
Female 50	Polyp high up (Side unknown)	Epistaxes	Repeated excision	Cure	11 years	Lack *et al.* (1977)
Female 50	Right middle turbinate	?	?	?	?	Lack *et al.* (1977)
Male 8	Left middle turbinate	Facial mass	Single excision	Cure	8 years	Lack *et al.* (1977)
Female 50	Right ethmoid complex	Endocrine upset	Radiotherapy	Symptom relief	6 years	Apple and Kreines (1982)
Female 31	Right ethmoid complex	Obstruction Discharge	Repeated excision	Cure	30 months	Veda *et. al.* (1985)
Female 56	Right inferior turbinate	Epistaxis	Radiotherapy Excision Radiotherapy	Symptom relief	21 years	Present case

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reported in Table I, to occur most often in middle-aged females and to present with local nasal symptoms. There is no record of metastatic disease in any of the cases. The most adequate treatment is total excision but this may involve considerable blood loss. Moran (1962) tied the right external carotid artery prior to the final curative excision and more recent authors have advocated embolisation prior to excision of a nasopharyngeal paraganglioma (Schuller and Lucas, 1982). Radiotherapy gave relief of symptoms in our case and in that of Apple and Kreines (1982) but did not cure the tumour in either case. Our patient is unusual in her extremely long follow-up, demonstrating the excellent response of the lesion to radiotherapy and local excision, despite failure to achieve complete cure. Her case also emphasises that, provided adequate records are available of the previous treatment, a second dose of radiation may yield good symptomatic relief a second time.

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