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# View from Beneath: Pathology in Focus

# Nasal haemangiopericytoma

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**Abstract**
Haemangiopericytoma is an uncommon vascular tumour frequently diagnosed with difficulty. The immunohistochemical findings of strong positivity to vimentin together with other diagnostic features (histological and ultrastructural) improves the certainty of its diagnosis. We report a case arising in the nose and outline diagnostic problems especially relating to histopathology.

## Introduction

Haemangiopericytoma is a rare vascular tumour arising from the proliferation of pericytes. By December 1988, only 24 cases of haemangiopericytoma arising from the nose and paranasal sinuses were reported in the literature. In 1990, however, Eichhorn *et al.*, reported eleven more cases, reassessing the electron microscopic findings and for the first time discussing the immunohistochemical features.

We report another case, bringing the total to 36 and we discuss the histopathological diagnosis based on the immunohistochemistry of sinonasal haemangiopericytomata.

## Case report

A 54-year-old white female, presented to the ENT outpatient clinic with a ten months history of left-sided nasal obstruction and a poor sense of smell. Examination confirmed reduced airflow through the left nostril but this was attributed to a deviated nasal septum to the left. She was listed for elective submucous resection of the nasal septum. On the operating table, examination of the nose showed a rather vascular polypoid lesion arising from the medial and posterior aspect of the left middle turbinate beyond the deflected nasal septum. The lesion was avulsed from its pedicle and sent for histology. Due to the suspicious nature of the lesion and the ensuing haemorrhage, the nose was packed and the operation abandoned. The nasal pack was removed after 48 hours with no further bleeding. The histology report was of haemangiopericytoma.

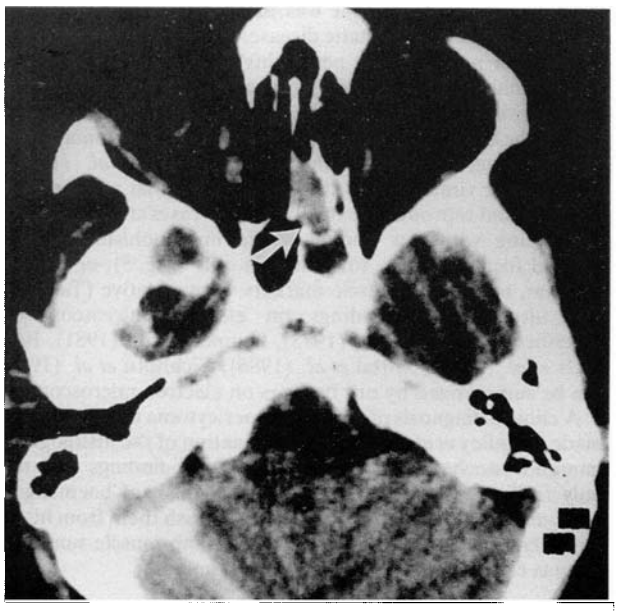

Fig. 1
CT scan of the nose and paranasal sinuses showing tumour.

The patient was readmitted for left middle turbinectomy but there was no sign of tumour. Three months later, there was evidence of tumour recurrence from the same site. A CT scan of the nose and paranasal sinuses, (Fig. 1), did not show evidence of a wide-spread lesion (*ie* intracranial involvement). The patient was readmitted and through a lateral rhinotomy incision, the left middle turbinate, including the tumour and a strip of septal mucosa, were excised. The patient has remained symptom free with no evidence of recurrence for the last nine months. She is still being followed up.

## Pathology

Histology showed a spindle cellular tumour, containing many ectatic vessels (Fig. 2). There was no invasion or destruction of the overlying mucosa. The cells were fusiform with little nuclear pleomorphism and very few mitotic figures; many cells showed vacuolation of the cytoplasm (Fig. 3). Mucin stains were negative, but reticulin was present around individual cells (Fig. 4). Immunohistochemistry showed focal positivity for vimentin only (Fig. 5), epithelial, vascular, neural and muscle markers were negative (Table I). Electron microscopy showed spindle and stellate cells lying outside the basal lamina of vessels. Intermediate sized filaments in bundles were present in the cytoplasm with mitochondria, some rough endoplasmic reticulin, and free ribosomes.

## Discussion

Ever since the first description of haemangiopericytoma by Stout and Murray in 1942, correct histopathological diagnosis is, at best, difficult and is made either with abandon by the unsuspecting or with extreme reluctance (Batsakis, 1979).

The submucosal location and intraluminal growth of these tumours, which are not encountered in haemangiopericytomata at other sites, makes them susceptible to ulceration of the surface epithelium with associated haemorrhage, inflammation and collagen deposition. These features might have led to errors in diagnosis (Eichhorn *et al.*, 1990).

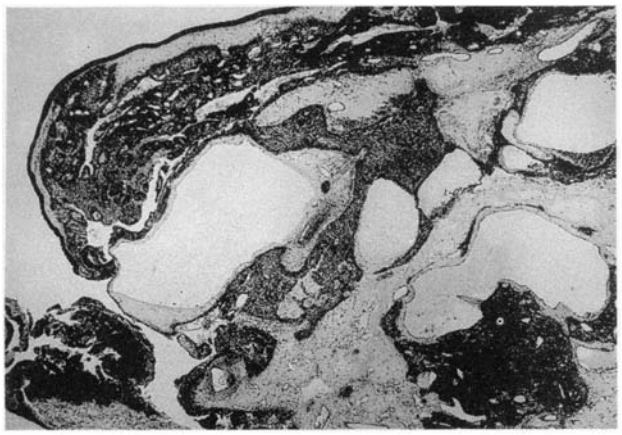

FIG. 2
Histology showing ectatic vessels with tumour in between.

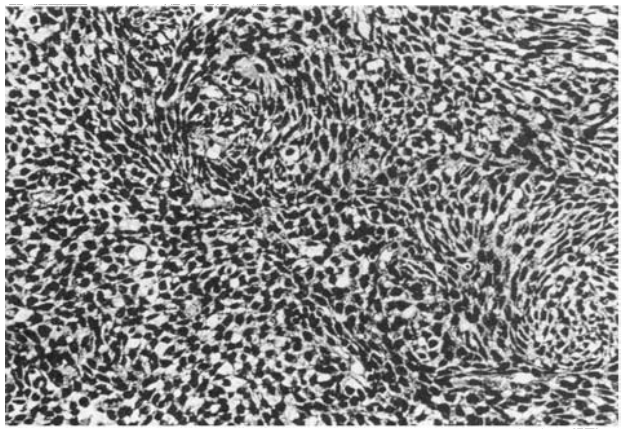

FIG. 3
Histology showing tumour cells with vacuolation of the cytoplasm

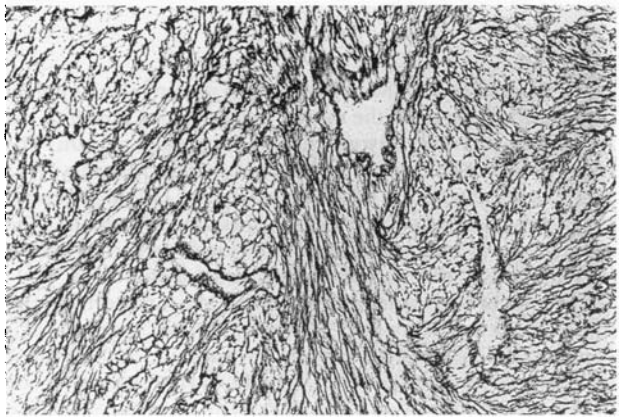

FIG. 4
Histology showing delicate reiculum network around individual cells.

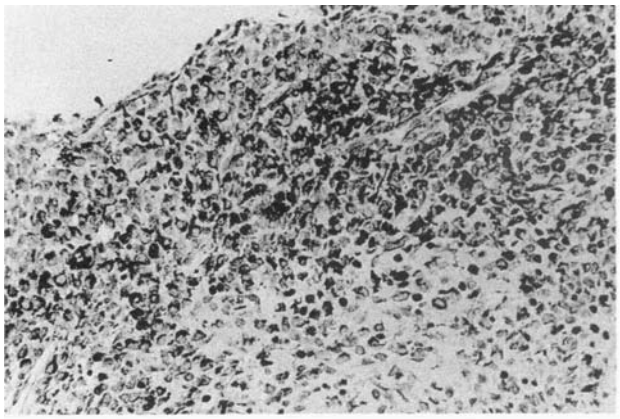

FIG. 5
Immunohistochemistry showing focal positivity for Vimentin.

Histopathologically, their variable vascular nature makes the distinction of haemangiopericytomata not always clear. This leads to the frequent difficulties encountered in arriving at a diagnosis made by the initial reviewers: haemangioblastoma (Stout and Murray, 1942), haemangioendothelioma (Ramsey, 1966), angioblastic meningioma (Pitkethly *et al.*, 1970), vascular sarcoma and haemangiopericytoma-like tumours (Compagno and Hyams, 1976).

Compagno and Hyams (1976) enumerated the cellular characteristics and nuclear features, which aid in making the diagnosis. They include absence of or minimal mitotic activity, clear distinction of normal vessels from tumour cells (usually by a delicate connective tissue sheath), uniform spindle cells with little or no overlapping of cellular borders, absence of necrosis and the presence of scattered mast cells. Enzinger and Smith (1976) suggest that the distinction between benign and malignant variants cannot be made in all cases, but emphasized the following features that was more often associated with recurrence and or metastatic disease: prominent mitotic activity (four or more mitosis per 10 high power field), necrosis, haemorrhage, increased cellularity and large size (>6.5 cm). In line with these characteristics, the histology in our case supported the diagnosis of a benign haemangiopericytoma.

TABLE I
RESULTS OF IMMUNOHISTOCHEMISTRY

| Antigen | Positivity |
|---|---|
| Cam 5.2 | – |
| Keratin | – |
| EMA | – |
| Factor VIII-related antigen | – |
| Chromogranin | – |
| NSE | – |
| S-100 | – |
| Desmin | – |
| Myoglobin | – |
| Actin | – |
| Vimentin | Focal +ve |

Immunoperoxidase staining by Eichhorn *et al.* (1990), showed that vimentin was the only antigen that was strongly detected and reproducible in all of the 10 cases studied. This is in keeping with our finding where immunohistochemistry showed focal positivity for vimentin only (Fig. 5); epithelial, vascular, neural and muscle markers were negative (Table I). The ultrastructural findings on electron microscopy as described by Hahn *et al.*, (1973), Nunnery *et al.* (1981), Batsakis *et al.* (1983), Mittal *et al.* (1986), Eichhorn *et al.* (1990) can be summarized by our findings on electron microscopy.

A clinical diagnosis of haemangiopericytoma has never been made (Bradley *et al.*, 1989). A combination of the histological, immunohistochemical, and ultrastructural findings will not only facilitate the histopathological diagnosis of haemangiopericytomata, but will also help to distinguish them from histologically similar neoplasms, such as smooth-muscle tumours, glomus cell tumours and schwann cell tumours.

**Acknowledgement**

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