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Haemangiopericytoma of the Parotid Gland

Report of a Case and Review of the Literature

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Summary. A primary malignant haemangiopericytoma of the parotid gland is reported. Microscopically the tumour showed two different types of area: in the first a histologically benign haemangiopericytoma was present, in the second, separated from the former by a clear-cut border, the structure was that of a histologically malignant haemangiopericytoma which subsequently metastasized and killed the patient.

The report stresses the great rarity of this neoplasm in the parotid gland and discusses the problem of the behaviour of haemangiopericytomas.

Key words: Haemangiopericytoma – Parotid neoplasms.

Introduction

Haemangiopericytoma is a rare neoplasm of the soft tissue first recognized by Stout and Murray in 1942. According to them the tumour has generally been regarded as deriving from Zimmerman's pericytes or, more correctly, as mimicking their morphological appearance.

The tumour is variable in its behaviour. The percentage of malignant haemangiopericytomas varies widely in different reviews ranging from 11.7% in Stout's report (1943) to 56.5% in O'Brien and Brasfield's paper (1965).

An apparent discrepancy between histology and behaviour has been said to be characteristic of this tumour. According to Stout (1949) "in most instances, it does not seem possible to distinguish between the benign and malignant by the histological appearance". However, further reports (Kauffman and Stout, 1960; Stout and Lattes, 1967; Enzinger et al., 1969) stress the existence of histologically recognizable benign and malignant forms. Moreover, McMaster et al. (1975) found a close relationship between histological appearance and

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biological behaviour. The tumour is ubiquitous in the body. A recent survey (Enzinger and Smith, 1967) reports that the lower extremities, retroperitoneum, head and neck are the most frequent sites of occurrence.

Connective tissue tumours of the major salivary glands are very rare, with the exception of haemangioma and lipoma. Foote and Frazell, (1954) stated that there was a need for publication of single case reports of such tumours. Haemangiopericytoma in the parotid gland is extremely rare; no mention of its occurrence is found even in detailed surveys (Thackray and Sobin, 1972; Thackray and Lucas, 1974).

To our knowledge only five cases of this tumour type have been recorded in the literature (Pellegrini et al., 1967; Cernea et al. 1969; Leonardelli and Bergomi, 1970; Hubert et al., 1970; Neal and Starke, 1973). Other cases may have been identified but the precise localization was not specifically stated as in the case of O'Brien and Brasfield (1965) which is depicted as situated in the 'parotid area'.

We observed a case of haemangiopericytoma of the parotid gland in which both benign and malignant areas were found to be present. The biological behaviour of this tumour was malignant.

Case Report

A 64 year old man was admitted to the Otolaryngology Department of Sassari University complaining of a painless mass in the left parotid region. The patient explained that the nodule had been present for about fifty years, since he was 14 years old. During the last two months, however, he had observed that the mass was rapidly growing.

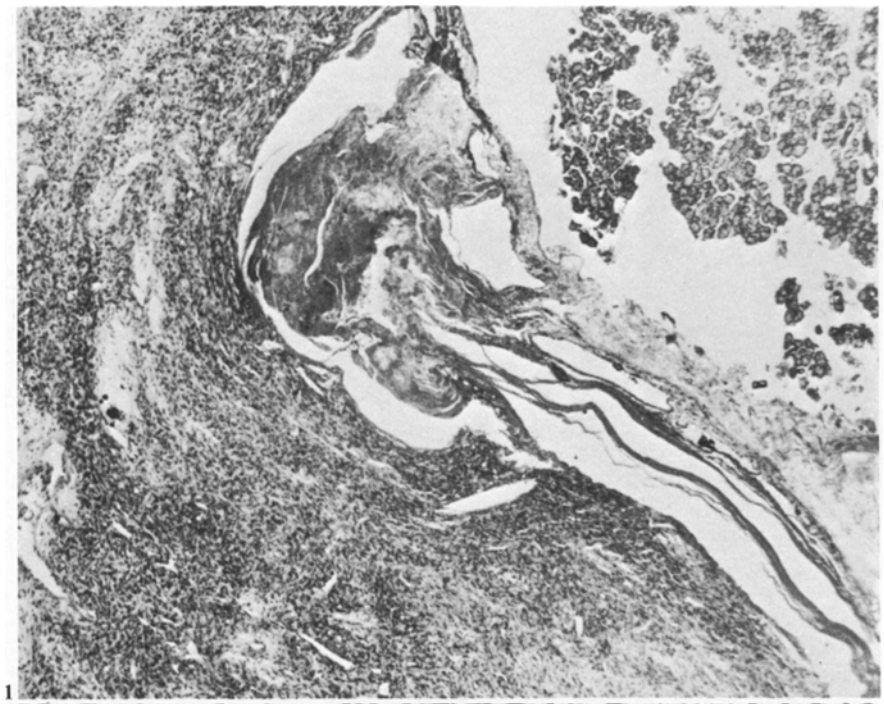
Physical examination to the left parotid gland showed a round mass of about 6.5 cm in diameter, firm in consistency and covered by stretched skin which was partially adherent to it. No deficit of the 7th cranial nerve or regional lymph node enlargement were found.

Laboratory tests were within the normal ranges. Sialography showed that the major ducts were compressed and interrupted by a mass growing in the postero-inferior area of the gland.

The patient underwent surgery for a suspected malignant tumour. During excision a frozen section was requested for pathological examination. A diagnosis of a probably malignant haemangiopericytoma was made. The parotid gland was removed but the 7th cranial nerve was preserved. Neck dissection was also performed.

Microscopical examination of paraffin sections from the whole surgical specimen ($6.5 \times 6 \times 5$ cm) showed a tumour well enveloped by a thick fibrous capsule and composed of round, oval or spindle-shaped cells with plump nuclei. Areas of stromal hyalinization were also visible (Fig. 1). A prominent feature was the presence of numerous, irregular star-like clefts layers by a single film of endothelial cells and containing a few RBCs: these were sinusoidal or capillary blood vessels (Fig. 2).

Silver staining for reticulin revealed a distinct basement membrane around these vessels with reticular fibres running radially along the proliferating cells. Due to their constant position as a mantle at the periphery of capillary blood vessels these cells were interpreted as pericytes. Mallory's phosphotungstic acid-haematoxylin (PTAH) stain showed no myofibrillar structures in their cytoplasm; Mallory's trichrome and Van Gieson's stains showed a weak positivity for collagen tissue. The periodic acid-Schiff (PAS) method was also negative.



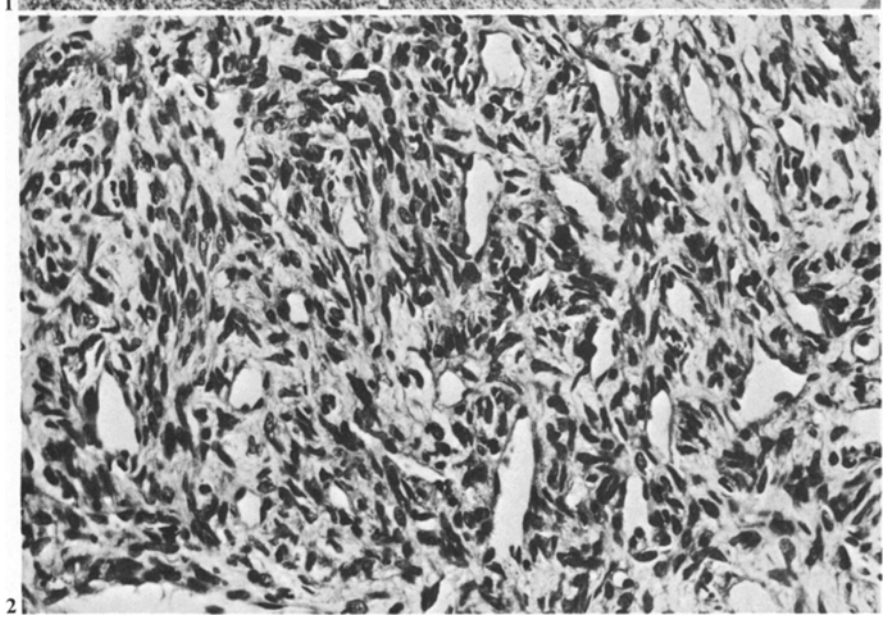


Fig. 1. A thick fibrous capsule separates the parotid gland tissue from the tumour. This is composed of spindle cell proliferation and contains many vascular spaces. Areas of stromal hyalinization are also visible. (Mallory's trichrome stain ×40)

Fig. 2. The vascular channels are surrounded by elongated proliferating cells. The vessels possess an easily distinguishable endothelial layer. The picture is compatible with a diagnosis of histologically benign haemangiopericytoma (H. & E. ×250)

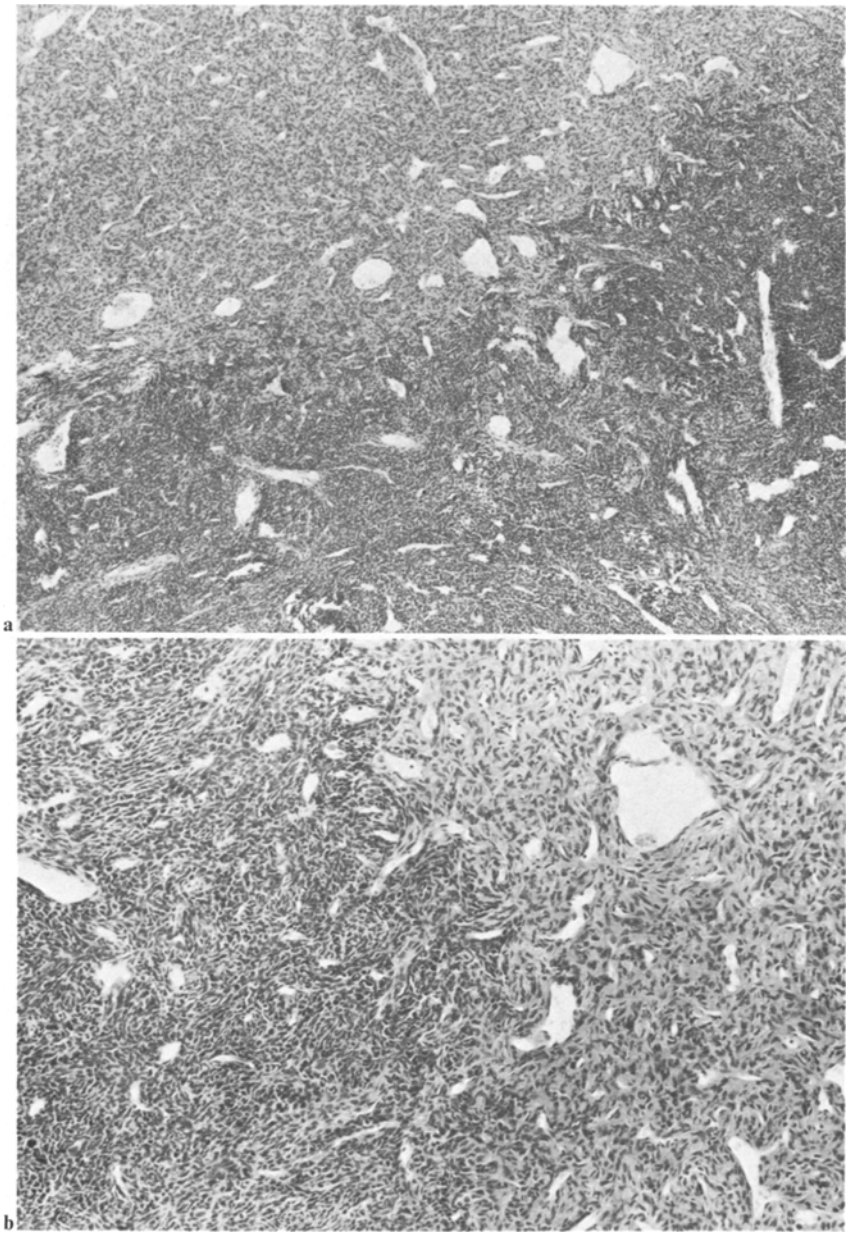


Fig. 3. a Two distinct types of appearance in the tumour are seen above and below a diagonal line. (H. & E. ×60). **b** In the first area (*right side*) a pattern of histologically benign haemangiopericytoma is recognizable. In the second area (*left side*) an increase in cell population with hyperchromatic nuclei and mitoses can be appreciated. The two areas are clearly merging. (H. & E. ×125)

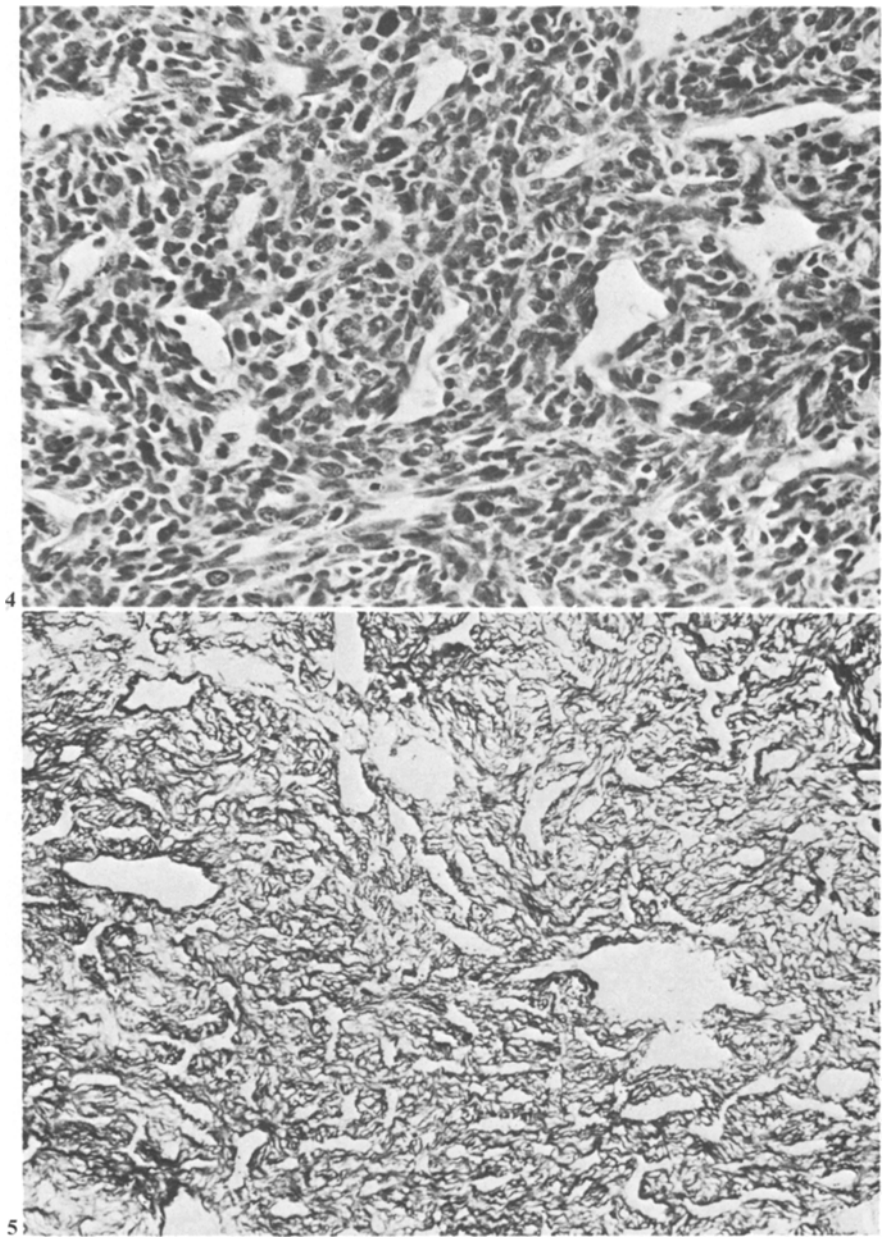


Fig. 4. The cells in a malignant area show many mitoses and have plump nuclei. Capillary blood vessels are prominent and still possess their endothelial layer. (H. & E. ×250)

Fig. 5. Silver stain for reticulin from a malignant part of the tumour reveals numerous vessels with a distinct basement membrane. The reticular fibres run radially along and around the proliferating cells (Gomori's method ×250)

No mitoses were present. A diagnosis of a histologically benign haemangiopericytoma was made.

However, in another area of the tumour, separated from the former by a clear-cut border, an increase in cell population could be observed (Fig. 3a, b). The cells were densely packed and showed hyperchromatic nuclei with numerous mitoses (10/hpf) (Fig. 4). Areas of hyalinization were no longer visible. The general appearance was that of a sarcomatous proliferation. The capillary blood vessels were less abundant, but still possessed their endothelial layer with a distinct basement membrane when silver stained (Fig. 5). Such features supported the diagnosis of a histologically malignant haemangiopericytoma, previously made on frozen sections.

No metastases to lymph nodes or invasion of the neighbouring soft tissues were found microscopically. The final diagnosis was "haemangiopericytoma of the parotid gland with malignant transformation".

The patient was dismissed with no further therapy other than surgical treatment, as suggested in the literature (Kauffman and Stout, 1960; O'Brien and Brosfield, 1965; Backwinkel and Diddams, 1970). He did well for 11 months when paresthesias and low back pain supervened. X-ray examination of the spine revealed a narrowing of the intervertebral space between D7–D8 due to pathological fracture. Moreover, myelography disclosed a block at the D10 level. Chest x-ray showed numerous metastatic nodules in both lungs. The patient was treated with chemotherapy and died 5 months later. No autopsy was performed.

Discussion

The tumour reported here is characterized by the following histological findings: a thick fibrous capsule, proliferation of oval or spindle-shaped cells around serpentine capillary blood vessels, absence of endothelial cell proliferation within the lumina, a distinct basement membrane surrounding capillary blood vessels and reticular fibres running from the basement membrane to the proliferating cells. Such features are consistent with a diagnosis of haemangiopericytoma (Stout, 1949; Begg and Garret, 1954; Enzinger and Smith, 1970).

In Table 1 we have listed the most important features of the cases of haemangiopericytoma of the parotid gland reported in the literature together with our own. Briefly the tumour affects males exclusively; the age incidence is between 20–23 years of age; the two older patients had had the tumour for many years. The tumour usually exceeds 3.5 cm in diameter and almost invariably exhibits malignant features.

In our case, in particular, two different areas of cell proliferation were present. *In the first* the spindle cells showed no mitoses, capillary blood vessels were clearly distinguishable and the stroma showed hyalinized areas, thus justifying a diagnosis of histologically benign haemangiopericytoma. In our opinion this diagnosis is correct although a spindle cell proliferation strictly related to blood vessels may simulated a vascular leiomyoma. However, such a possibility was excluded because of the characteristic pattern of the reticulin network in our case. Glomus tumour, another neoplasm of pericytes (Stout, 1956), was

Table 1. Haemangiopericytomas of the parotid gland. Summary of data collected by authors

Author	Sex	Age	Site	Duration of symptoms	Size (cm)	Histology	Follow up	Metatases
Pellegrini (1967)	M	20	left	1 month	5×3	malignant	alive 2 months after diagnosis	not known
Cernea (1969)	M	23	right	6 months	4×2	benign	alive 5 months after diagnosis	not known
Hubert (1970)	M	21	left	4 months	not known	malignant	died 32 months after diagnosis	lungs, bone
Leonardelli (1970)	M	57	right	10 years	3.5×3	malignant	alive 29 months after diagnosis	not known
Neal (1973)	M	21	left	3 years	8×5×6	malignant	died 38 months after diagnosis	lungs, bones
Present case	M	64	left	50 years	6.5×6×5	malignant	died 16 months after diagnosis	lungs, bones

not considered in the differential diagnosis because of the absence of organoid structures.

In the second area the spindle cells were crowded and showed many mitoses. Capillary blood vessels were less prominent, but retained their basement membrane and endothelial cells. Hence a diagnosis of histologically malignant haemangiopericytoma was justifiable (Kauffman and Stout, 1960; McMaster et al., 1975). This sarcoma-like appearance makes the differential diagnosis more intriguing. In fact a diagnosis of synovial sarcoma, fibrosarcoma or Kaposi's sarcoma (fibroblastic stage) might be suggested. In our experience, however, a silver stain for reticulin showing a distinct basement membrane excludes synovial sarcoma (Massarelli et al., 1978). Moreover, the reticular fibres around the proliferating cells are not consistent with a fibrosarcoma in which reticulin does not show a distinctive arrangement and capillary blood vessels are very inconspicuous (Ashley, 1978). Finally, Kaposi's sarcoma could be excluded because erythrocytes are not contained in capillary blood vessels in the fibroblastic stage, but are dispersed throughout the fibroblastic proliferation (Lever and Schaumburg-Lever, 1975).

Apart from its unusual localization the present case shows a few aspects that are worth discussing. The patient mentioned that the tumour had been present for fifty years, beginning at the age of 14. The age of onset and the long benign biological behaviour could support the view that the tumour was initially of the juvenile type. According to Kauffman and Stout (1960) and Enzinger and Smith (1976) this variant is believed to be less malignant than the adult form.

Further, the occurrence of both benign and malignant areas in the same tumour suggests that the neoplasm remained benign for many years and became malignant late in its natural history. Actual malignancy of the tumour was supported by the histological picture. McMaster et al. (1975) found a close

relationship between histology and behaviour partially modifying Stout's statement about the difficulty in assessing the prognosis on histological grounds. In our opinion, the apparent contradiction can be explained by the difficulty in detecting small foci of malignancy in tumours which are otherwise histologically benign (McCormack and Gallivan, 1954). The malignancy of the tumour was confirmed by metastases to the lungs and bones and by the sparing of regional lymph nodes, as reported in the literature (Stout, 1949; O'Brien and Brasfield, 1965; McCormack and Gallivan, 1954).

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