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Haemangiopericytoma of the parotid salivary gland: report of a case with literature review

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Abstract
Haemangiopericytoma is a rare, soft tissue, tumour with unpredictable biological behaviour. A case of haemangiopericytoma of the parotid salivary gland is reported. The clinical, surgical and histological features are described.

Key words: Salivary gland neoplasms; Haemangiopericytoma; Parotid gland

Introduction

Haemangiopericytoma was first described by Stout and Murray (1942). The neoplasm was characterized as being composed of capillaries surrounded by an accumulation of small, oval, cells which Zimmermann (1923) had called pericytes. Haemangiopericytomas may arise wherever capillaries are found, although they usually occur in the soft tissues of the extremities i.e. retroperitoneum, pelvic fossa and head and neck (Stout, 1956; O'Brien and Brasfield, 1965; Backwinkel and Diddams, 1970; Enzinger and Smith, 1976). According to Batsakis and Rice (1981) its incidence in the head and neck lies between 15 per cent and 25 per cent of all haemangiopericytomas.

Haemangiopericytoma is a rare, vascular, tumour which represents only 1.3 per cent of all vasoformative neoplasms of the body (Walike and Bailey, 1971) approximately 50 per cent of the reported cases being malignant (Backwinkel and Diddams, 1970). Although there have been reports of a traumatic association (Stout, 1949) and steroid therapy (Masson *et al.*, 1950) the aetiology of these tumours is still obscure. There does not appear to be any relationship with systemic disturbances and these tumours in the head and neck region, although there have been reports of hypoglycaemia due to glucose hyperutilization (Howard and Davis, 1959) and hypertension related to a renin-secreting tumour (Robertson *et al.*, 1967) causing systemic disturbances possibly related to these tumours.

Stout (1956) reported 197 cases of haemangiopericytomas of which one involved the salivary gland. Pagliaro *et al.* (1988) on reviewing the literature reported that 18 cases of haemangiopericytomas of the major salivary glands had been recorded, of which 16 were found in the parotid gland, (O'Brien and Brasfield, 1965; Pellegrini *et al.*, 1967; Cernea *et al.*, 1969; Farr *et al.*, 1970; Hubert *et al.*, 1970; Leonardelli and Bergomi, 1970; Gerner *et al.*, 1973; Neale and Starke, 1973; Yamaguchi *et al.*, 1977; Peynegrie and Pain, 1978; Massarelli *et al.*, 1980; Auguste *et al.*, 1982; Bertrand *et al.*, 1984; Baicco *et al.*, 1985; Tatum *et al.*, 1986). More recently Phillippou and Gellrich (1992) reported a further case coupled with the case just presented which makes a total of 18 cases.

Haemangiopericytoma is a primary tumour of adults and rare in children (Enzinger and Weiss, 1988). Tumours occur equally in the sexes. Most patients with this tumour complain of an enlarging painless mass and many reach a fairly large size before help is sought. The case presented for discussion had a history of 22 years of swelling in the right parotid region and only in the last twelve months had the swelling caused any discomfort. Because of the rich vascularity, telangiectasia and raised temperature of the overlying skin may occur, as well as pulsation and an audible bruit.

In the series by Enzinger and Smith (1976) reporting 106 cases, 17 were found to be in the head and neck region. The haemangiopericytoma-like tumours of the nasal passages and paranasal sinuses differ slightly from these elsewhere in the body and may represent a related but separate entity (Campagno and Hyams, 1976; Campagno, 1978).

Case report

E.B. an 82-year-old lady first presented to the ENT Department in September 1989. She gave a 22-year history of a cystic swelling in the right parotid region which had noticeably got bigger in the last two years. She described an occasional burning discomfort in the right side of her face, and also an increasing tight feeling. She did not describe any facial weakness. She had no constitutional symptoms.

Examination revealed a 6 × 3 cm smooth but lobulated swelling in the right parotid region, which was immobile but not attached to skin (Figure 1). There was no evidence of facial weakness. There was no palpable cervical lymphadenopathy. She was noted to have a right carotid bruit. A CT scan (Figure 2) confirmed the mass in the region of the right parotid gland. It appeared to originate in the gland itself, but the normal glandular tissue was displaced inferiorly and medially by the lesion. Following intravenous (IV) contrast there was very dense but not entirely homogenous enhancement. The appearances were those of a haemangioma which extended into the deep part of the parotid.

At that time the patient declined surgical excision and was seen at regular intervals in the Outpatients Clinic. Some eighteen months later the swelling increased rapidly in size and was causing the patient discomfort, particularly at night.

The mass was excised with no damage to the facial nerve. Post-operatively there was a collection of blood-stained fluid aspirated from under the skin flap but the remainder of her post-operative course was uneventful. A year later the patient was fit and well with no evidence of recurrent tumour (Figure 3).

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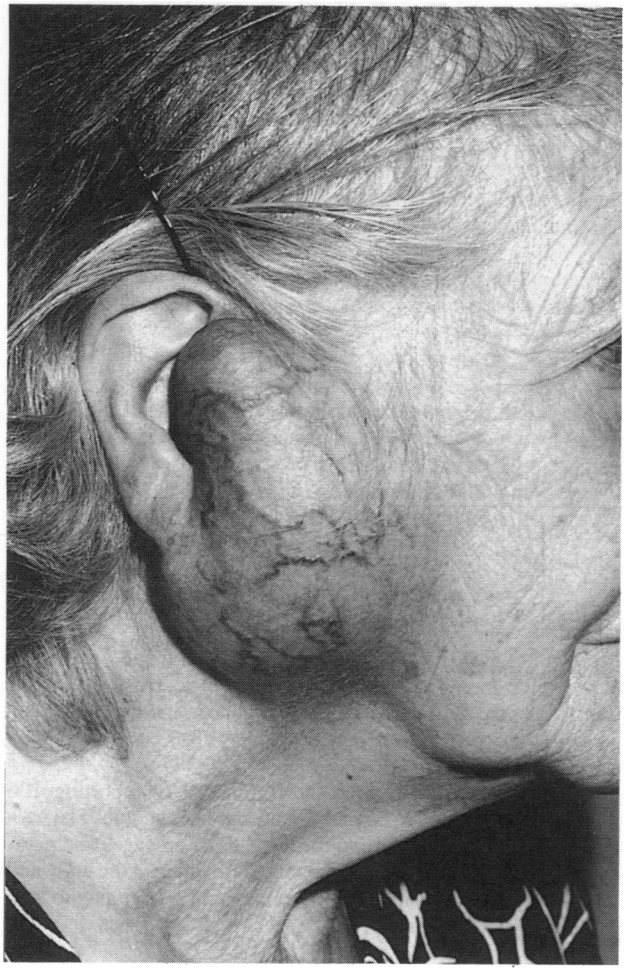

FIG. 1
Presenting features of the parotid mass.

Pathology

Gross appearances

The specimen was an irregularly-lobulated mass measuring 90 × 50 × 30 cm and weighing 70 gm. On slicing it, part of the lesion was found to be solid, but much of it was cystic with a translucent appearance and jelly-like consistency.

Microscopy

Sections showed a tumour composed of numerous thin-walled vascular channels between which were tightly-packed, regular, ovoid cells which had fewer than two mitotic figures per 10 high power fields (HPF). Focally there was a slight storiform pattern and in part of the lesion large degenerative cyst-like spaces were seen with some surrounding mucoid matrix (Figure 4).

Immunostaining for S100 and epithelial markers was negative and there was no stainable glycogen. No haemorrhage or other features of aggressive behaviour were present. The features of a spindle-cell tumour, with closely investing, arborizing, thin-walled blood vessels were characteristic of a haemangiopericytoma.

Discussion

The problem of reaching an accurate diagnosis and predicting the clinical behaviour of these tumours has been stressed in the literature and there is considerable variation in the reporting of benign and malignant haemangiopericytoma. A report of 106 cases by Enzinger and Smith (1976) showed that benign tumours outnumbered malignant ones by a sizeable margin. Their criteria

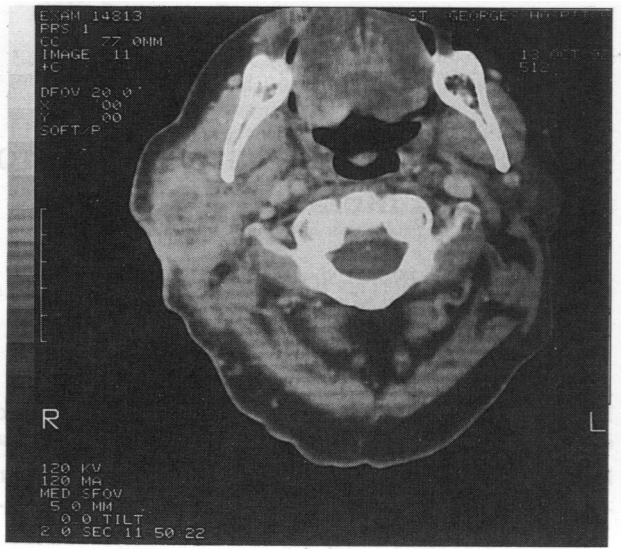

FIG. 2
CT scan (R) parotid region.

for differentiating between benign and malignant tumours, namely a tumour diameter of 6.5 cm, prominent mitotic activity (four or more mitotic figures per 10 HPF) haemorrhage, necrosis, cellular anaplasia and increased cellularity associated with thrombosis can lead to a more accurate diagnosis. Tumour size also appears to play a role, but there is no evidence that the anatomical site of the tumour significantly affects its behaviour.

A review of 60 cases by McMaster *et al.* (1975) used slightly more stringent criteria for diagnosis. They stated that a slight degree of anaplasia with one mitotic figure per 10 HPF or moderate degree of cellular anaplasia and one mitotic figure per 20 HPF would be more likely to lead to malignant behaviour. Reported rates of metastasis vary from 11.7 per cent (Smullens *et al.*, 1982) to 56.5 per cent (O'Brien and Brasfield, 1965). The lung and skeleton are the more frequent metastatic sites. Lymph node metastasis is rare. Enzinger and Smith (1976) report one case out of 106 cases.

Recurrence at the original site is an ominous sign, the majority of recurrent tumours develop metastasis at a later date (Enzinger and Smith, 1976).

These tumours have been known to recur years or even decades later. Shin *et al.* (1987) and Rice *et al.* (1989) reported cases recurring 28 and 33 years later respectively. A period of at least 10 years has been suggested for follow-up (Phillippou and Gellrich, 1992), they also considered all haemangiopericytomas as potentially malignant, an isolated view not previously reported in the literature.

FIG. 3
Post-operative appearance.

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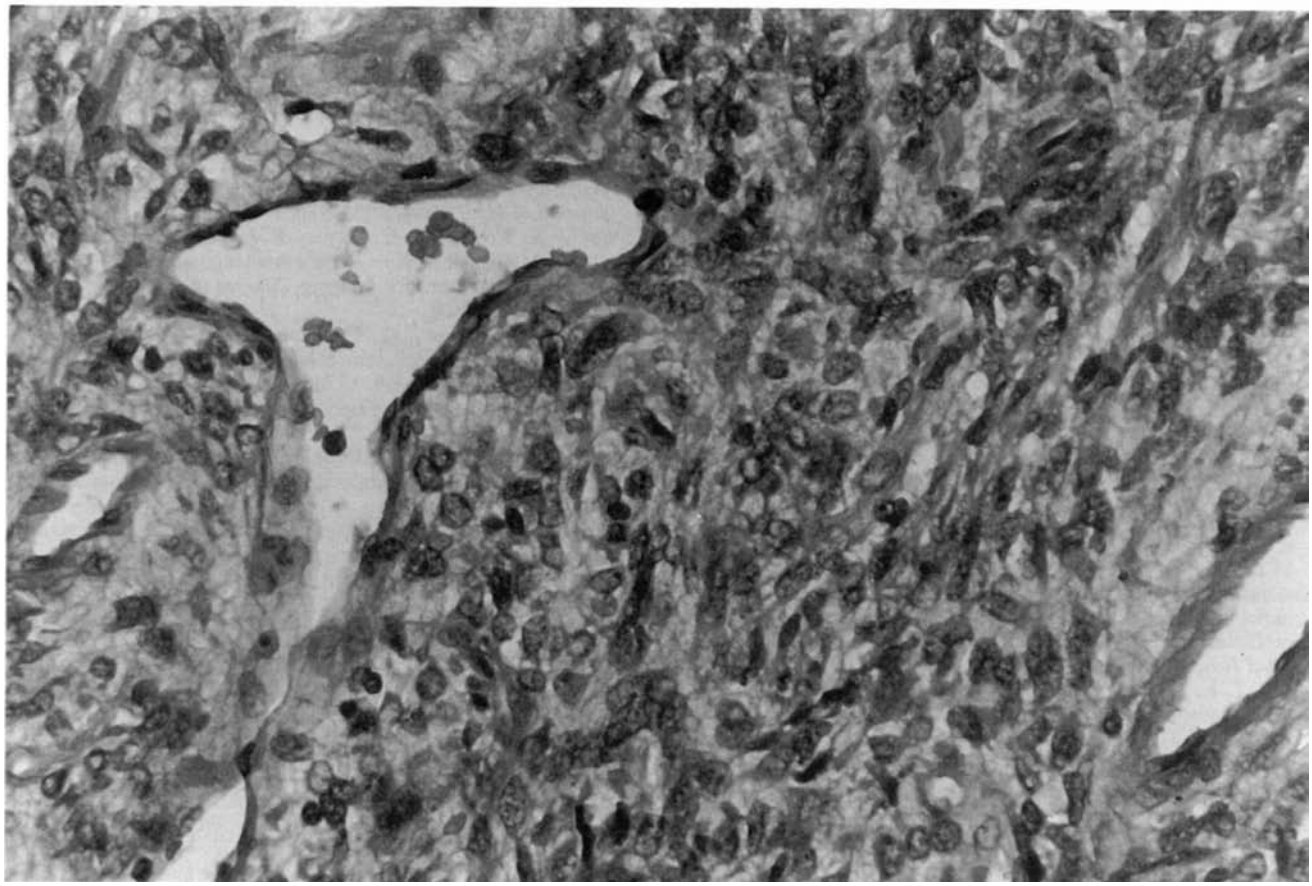

FIG. 4

Histological appearance of haemangiopericytoma. (H & E ×400).

It is generally accepted that the treatment of choice is wide surgical excision. Cases treated by surgery alone show a cure rate of 53.1 per cent compared to a 13.3 per cent cure rate for radiotherapy although radiotherapy is recommended at least in cases of incomplete tumour removal (Backwinkel and Diddams, 1970). Surgical removal may be facilitated if the afferent vessels are ligated during the initial stage of the operation. Mira *et al* (1977) reported satisfactory response to pre-operative embolization, surgical removal and radiation with complete regression in 47 per cent of the cases. Response was best with doses over 35 Gy and tumours measuring less than 5 cm. Smullens *et al.* (1982) also recommended pre-operative embolization. The use of chemotherapy has had a mixed response as reported in the literature. Positive response have been reported for adriamycin alone or in combination with cyclophosphamide, vincristin, methotrexate or actinomycin D (Ortega *et al.*, 1971; Wong and Yagoda, 1978; Beadle and Hillcoat, 1983; Nathanson, 1984), although some authors consider haemangiopericytomas to be chemotherapy resistant (McMaster *et al.*, 1975; Atkinson *et al.*, 1984; Weber *et al.*, 1990).

Summary

Haemangiopericytoma is a rare, vascular, soft tissue, tumour with a variable pathological behaviour.

Wide surgical treatment is the treatment of choice, but radiotherapy and chemotherapy may be used as adjuncts.

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References

Atkinson, J. B., Mahoor, G. H., Isaacs, H., Ortega, J. A. (1984) Haemangiopericytoma in infants and children. *American Journal of Surgery* **148:** 372–374.

Auguste, L. J., Razack, M. S., Sako, K. (1982) Haemangiopericytoma. *Journal of Surgical Oncology* **21:** 260–264.

Backwinkel, K. D., Diddams, J. A. (1970) Haemangiopericytoma: report of a case and comprehensive review of the literature. *Cancer* **25:** 896–900.

Baicco, F., Rognoni, S., Negri, A. (1985) Emangiopericitoma parotideo. Descrizione di un case. *Il Valsaval* **61:** 123–126.

Batsakis, J. G., Rice, D. H. (1981) The pathology of head and neck tumours: vasoformative tumours, part 9B. *Head and Neck Surgery* **3:** 326–338.

Beadle, G. F., Hillcoat, B. L. (1983) Treatment of advanced malignant haemangiopericytoma with combination adriamycin and DTIC: a report of four cases. *Journal of Surgical Oncology* **22:** 167–170.

Bertrand, J. C., Guilbert, F., Vaillant, J. M., Szpirglas, H., Chomette, G., Auriol, M., Mercier, C. (1984) Les hemangiopericytomes de la sphere orofaciale. *Annales d'Otoloryngologie et de Chirurgie Cervico-Faciale* **101:** 607–613.

Campagno, J. (1978) Hemangiopericytoma-like tumours of the nasal cavity: a comparison of haemangiopericytoma of soft tissue. *Laryngoscope* **88:** 460.

Campagno, J., Hyams, J. (1976) Hemangiopericytoma-like intranasal tumours. A clinico-pathologic study of 23 cases. *American Journal of Clinical Pathology* **66:** 672.

Cernea, P., Debry, D., Lavdenbach, P., Moreau, R., Brocheriou, C., Vaillant, J. M., Guilbert, F., Soubiran, J. M., Chevallier, C., Hosxe, G., Peri, G. (1969) Haemangiopericytome de la parotide. *Revue de Stomatologie et de Chirurgie Maxillo-Faciale* **70:** 132–135.

Enzinger, F. M., Smith, B. H. (1976) Haemangiopericytoma. An analysis of 106 cases. *Human Pathology* **7:** 61–82.

Enzinger, F. M., Weiss, S. W. (1988). *Soft tissue tumors*. 2nd Edition. Ch. 23. C. V. Mosby Co., New York, pp 596–613.

Farr, H. W., Carandang, C. M., Huvos, A. G. (1970) Malignant vascular tumours of the head and neck. *American Journal of Surgery* **120:** 501–504.

Gerner, R. E., Moore, G. E., Pickren, J. W. (1973) Haemangiopericytoma. *Annals of Surgery* **177:** 128–132.

Howard, J. W., Davis, P. L. (1959) Retroperitoneal haemangiopericytoma associated with hypoglyacaemia and masculinization. *Delaware State Medical Journal* **51:** 29–34.
Hubert, J. C., Drevet, D., Gay, R. (1970) À propos d'un cas d'haemangiopericytoma parotidien. *Journal Francais d'Oto-Rhino-Laryngologie, Audiophonologie et Chirurgie Maxillo-Faciale* **19:** 659–664.
Leonardelli, G. B., Bergomi, A. (1970) Une tumeur rare de la parotide: l'hemangiopericytome de Murray et Stout. *Cahiers d'Oto-Rhino-Laryngologie et de Chirurgie Cervico-Faciale* **5:** 37–40.
Massarelli, G., Tanda, F., Fois, V., Oppia, L. (1980) Haemangiopericytoma of the parotid gland. Report of a case and review of the literature. *Virchows Archives A: Pathological Anatomy and Histology* **368:** 81–89.
Masson, G. M. C., Hazard, J. B., Corcoran, A. C. (1950) Experimental vascular disease due to desoxycortiosterone and anterior pituitary factors. II: Comparison of pathological changes. *Archives of Pathology* **49:** 641–664.
McMaster, M. J., Soule, E. H., Ivans, J. C. (1975) Haemantiopericytoma. A clinicopathologic study and long-term follow-up of 60 patients. *Cancer* **36:** 2232–2244.
Mira, J. G., Chu, R. C., Fatner, J. G. (1977) The role of radiotherapy in the management of malignant hemangiopericytoma. Report of 11 new cases and review of the literature. *Cancer* **39:** 1254.
Nathanson, D. S. (1984) Pre-operative chemotherapy and irradiation in the treatment of soft tissue sarcomas of the lower extremities; preliminary report of experience at Henry Ford Hospital. *Henry Ford Hospital Medical Journal* **32:** 31–38.
Neale, T. F., Starke, W. P. (1973) Haemangiopericytoma of the parotid gland: a case report with autopsy. *Laryngoscope* **83:** 1953–1958.
O'Brien, P., Brasfield, R. D. (1965) Haemangiopericytoma. *Cancer* **18:** 249–252.
Ortega, J. A., Finklestein, J. Z., Hittle, R. (1971) Chemotherapy of malignant haemangiopericytoma of childhood. *Cancer* **27:** 730.
Pagliaro, G., Poli, P., Ralza, G., Grandi, G. (1988) Haemantiopericytoma of the sub-mandibular gland: case report. *Journal of Laryngology and Otology* **102:** 97–99.
Pellegrini, G., Verdaguer, S., Boyre, F., De Firmas, J. L., Philip, F. (1967) Haemangiopericytome parotidien. *Revue de Laryngologie, Otologie, Rhinologie* **28:** 421–427.
Peynegrie, R., Pain, F. (1978) Un haemangiopericytome de la parotide chez l'enfant. *Anales d'Oto-Laryngologie et de Chirurgie Cervico-Faciale* **95:** 789–793.
Phillippou, S., Gellrich, N. C. (1992) Haemangiopericytoma of the head and neck region. A clinical and morphological study of three cases. *International Journal of Oral Maxillo-facial Surgery* **21:** 99–103.
Rice, C. D., Kersten, R. C., Mrak, R. E. (1989) An orbital haemangiopericytoma recurrent after 33 years. *Archives of Ophthalmology* **107:** 552–556.
Robertson, P. W., Klidjian, A., Hardin, L. K., Walters, G. (1967) Hypertension due to resin-secreting renal tumour. *American Journal of Medicine* **43:** 963–976.
Shin, M. S., Koehler, R. E., Stanley, R. J., Barton, J. C., Ho, K. J. (1987) Malignant haemangiopericytoma; computed tomography and magnetic resonance imaging. *Journal of Computer Tomography* **11:** 297–300.
Smullens, S. N., Scotti, D., Osterholm, J. L. (1982) Preoperative embolization of retroperitoneal haemangiopericytomas as and in their removal. *Cancer* **50:** 1870.
Stout, A. P. (1949) Haemangiopericytoma. A study of 25 new cases. *Cancer* **2:** 1027–1054.
Stout, A. P., Murray, M. R. (1942) Haemangiopericytoma: vascular tumour featuring Zimmerman's pericytes. *Annals of Surgery* **116:** 26–33.
Stout, A. P. (1956) Tumours featuring pericytes; glomus tumour and haemangiopericytoma. *Laboratory Investigation* **6:** 217–223.
Tatum, R. C., Skinner, H. C., Adrian, J. (1986) Haemangiopericytoma of the parotid: report of a rare case with literature review. *Quintessence International* **17:** 399–403.
Walike, J. W., Bailey, B. J. (1971) Head and neck haemangiopericytoma. *Archives of Otolaryngology* **93:** 345–353.
Weber, B. P., Sshrader, M., Inniger, R. (1990) Maligne Hamangiopericytome in Kopf. Hals-Bereich. *HNO* **38:** 326–333.
Wong, P. P., Yagoda, A. (1978) Chemotherapy of malignant haemangiopericytoma. *Cancer* **41:** 1256–1260.
Yamaguchi, K. T., Krugman, M. E., Barr, R. J., Reingold, I. M. (1977) Haemangiopericytoma of the parotid gland with a review of the literature. *Journal of Otolaryngology* **6:** 431–435.
Zimmermann, K. W. (1923) Der Feinere Ban de blut capillaren. *Zeitschrift für Anatomie und Entwicklungsgeschichte* **98:** 29–109.

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