J Clin Periodontol 2005; 32: 921–924 doi: 10.1111/j.1600-051X.2005.00783.x

Journal of Clinical Periodontology

Case Report

Haemangiopericytoma of the maxillary gingiva: report of a case

Giovanna Petrone[1], Vittoria Perrotti[1], Massimiliano Fioroni[2], Corrado Rubini[3] and Adriano Piattelli[1]

[1]Dental School, University of Chieti-Pescara, Chieti, Italy; [2]Dental School, University of Ancona, Ancona, Italy; [3]Department of Pathology, University of Ancona, Ancona, Italy

Petrone G, Perrotti V, Fioroni M, Rubini C, Piattelli A. Haemangiopericytoma of the maxillary gingiva: report of a case. J Clin Periodontol 2005; 32: 921–924. doi: 10.1111/j.1600-051X.2005.00783.x. © Blackwell Munksgaard, 2005.

Abstract

Aim: Haemangiopericytoma (HPC) represents approximately 3% of all tumours in the head and neck. This tumour is a soft tissue tumour derived from mesenchymal cells with pericytic differentiation. We present the clinicopathological findings of a case.

Materials and Methods: A 69-year-old man was referred to our Department for a mass located on the right pre-molar maxillary gingiva; this mass caused problems during chewing, but was otherwise asymptomatic.

Results: Clinical examination revealed a nodular, pink lesion, 3.5 cm in diameter, which was lined with normal mucosa. The lesion was mobile in relation to the deep and superficial tissues. Microscopic analysis of the neoplasm showed a vascular rich pattern, constituted by vessels covered with flat endothelium and surrounded by abundant spindly cells. On the basis of these histological and immunohistochemical findings, the final diagnosis was HPC.

Conclusions: HPC is an uncommon vascular tumour for which the biological behaviour is difficult to predict. In our patient, no recurrences or distant metastases were present at a 4 years follow-up.

Key words: gingiva; haemangiopericytoma; vascular tumour

Acceptance for publication 16 March 2005

Haemangiopericytoma (HPC) is rare and it represents about 1% of all adult vascular neoplasms (Sikes et al. 2003). The majority of HPCs occur in superficial cutaneous tissues or in muscle layers of the lower extremities, retroperitoneum and pelvis (Sikes et al. 2003).

Initially described by Stout (1942), HPC is a soft tissue tumour derived from mesenchymal cells with pericytic differentiation (Llorente et al. 1999, Enzinger & Weiss 2001, Alabdulhadi et al. 2004). These pericytes have uniform, ovoid nuclei with indistinct cell borders and are located outside the reticulin sheath of the endothelium. Although their precise function is unknown, these ovoid cells are intimately associated with capillaries and possess structural similarity to smooth muscle cells and fibroblasts, suggesting a contractile and supportive role (McMaster et al. 1975, Alabdulhadi et al. 2004). Sites involved by HPC include the trunk, skin and retroperitoneum. About 15% of all HPC occur in the head and neck; in this area the sinonasal tract is the most common site accounting for 55% of all cases (Barnes 2001). In the head and neck, HPC involves orbit, nasopharynx, parotid gland, tongue, larynx, trachea, thyroid, forehead, scalp, temporal bone, anterior skull, occiput, ear and periauricular area (Walike & Bailey 1971, August et al. 1982, Abdel-Fattah et al. 1990, Sellke et al. 1991, Carew et al. 1999 & Barnes 2001). HPC of the oral cavity has been observed in the floor of the mouth, hard palate, gingiva, alveolar-buccal sulcus, mandible and soft palate (Goldwasser & Daw 1990, Vogler et al. 1990, Singh et al. 1993, Hiraumi et al. 2002). HPC represents approximately 3% of all tumours in the head and neck (Walike & Bailey 1971). It can occur in any age group, but the most affected age is the fifth and sixth decades (Omer et al. 1995). Paediatric cases account for less than 10% of all HPC (Enzinger & Smith 1976). There is no sex predilection (McMaster et al. 1975, Enzinger & Smith 1976). The aetiology of HPC is still unknown, but an association with trauma has been suggested (Stout 1942).

We report a case of HPC located in the right maxillary gingiva.

Case Report

A 69-year-old man presented in our Department complaining of a mass, which had appeared a few months previously, located between the right maxillary adherent gingiva and alveolar mucosa in the pre-molar area (Fig. 1). No bleeding was present. Clinical examination revealed a nodular, pink, non-tender lesion, of about 3.5 cm diameter, with tense-elastic consistency, which was covered with normal, healthy, not ulcerated mucosa. The lesion was mobile in relation to the deep and superficial tissues. Radiologically, no erosion of the underlying bone was present. Both pre-molars were vital. The lesion caused problems during chewing, but

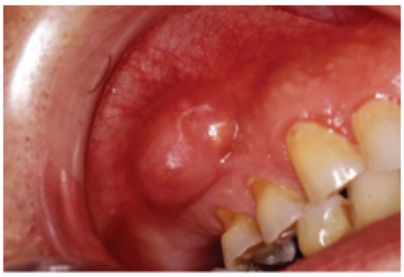

Fig. 1. Clinical appearance of the lesion at first presentation.

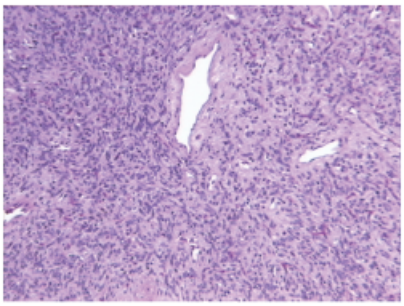

Fig. 2. Haemangiopericytoma. Microscopic analysis of the neoplasm showed a rich vascular pattern, constituted by vessels lined with flat endothelium surrounded by abundant spindly (pericytic) cells. H&E × 160.

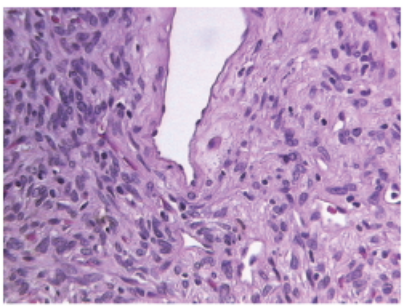

Fig. 3. Haemangiopericytoma. Hyalinization of collagen was present next to the perivascular area. H&E × 250.

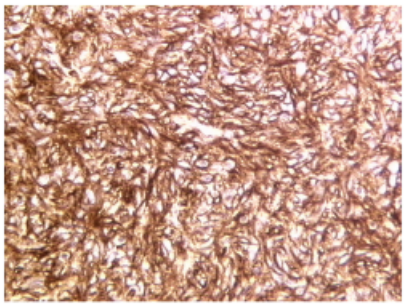

Fig. 4. Haemangiopericytoma. The cells were circumscribed by a dense reticulum. Reticulin staining × 160.

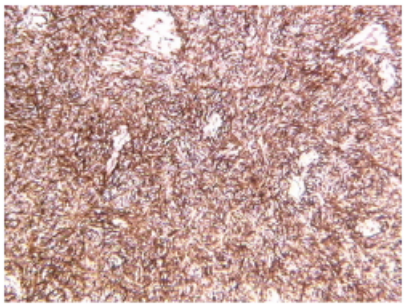

Fig. 5. Haemangiopericytoma. Immunohistochemically the tumour cells were intensely positive for CD34. CD34 (PaP) × 160.

was otherwise asymptomatic. Excisional biopsy of the lesion was performed with the patient under local anaesthesia, and the tissue was sent for histological evaluation. Histologically, the lesion showed a rich vascular pattern, constituted by vessels lined with flat endothelium (Fig. 2). Cell nests, constituted by relatively monomorphic cells that tended to be spindle shaped and arranged in bundles, were present. Hyalinization of collagen was present next to the perivascular area (Fig. 3). The cells were surrounded by a dense reticulum (Fig. 4). Immunohistochemically, the tumour cells were intensely positive for CD34 (Fig. 5) and for vimentin, and completely negative for S100 protein, smooth-muscle actin, desmin, cytokeratins and EMA. Mitotic figures were rare (1–210 high-power fields (HPF)) and there was focal evidence of cytologic atypia. On the basis of these histological and immunohistochemical findings, the final diagnosis was an HPC. Physical examination and a computerized tomography scan of the neck, liver and lungs ruled out the possibility of metastasis elsewhere in the body. The patient subsequently underwent a wide surgical excision of the lesion. No recurrences or distant metastases were present at a 4 years follow-up.

Discussion

HPC presents usually as a deep soft tissue mass with an insidious growth, and may be associated with pain (Walike & Bailey 1971, Kowalski & Paulino 2001). Clinically, this tumour has a grey–white or brownish cut surface and its size can range from 1 to 20 cm (Volpe et al. 1991). Macroscopically, HPC can be lobulated or nodular, firmly attached to muscle or fascia; it can be soft, spongy, firm or friable and has, usually, a well-defined capsule. Microscopically, it is characterized by a proliferation of variably sized pericytes and by branching, thin-walled vascular channels. Radiographical features are not specific: they consist of a well-circumscribed, radiopaque soft tissue mass that often displaces neighbouring structures, and can include osteolysis of the involved bone (Wold et al. 1982).

Microscopically, diagnostic difficulties may be present because the tumour cells are undifferentiated and resemble those seen in several other tumours (Kempson et al. 2001). Vimentin is the only marker that is consistently expressed in HPC (Barnes 2001, Sikes et al. 2003). Some tumours may be positive for smooth muscle actin (0–72%) or S-100 protein (0–14%) (Barnes 2001). Desmin, cytokeratins, epithelial membrane antigen and factor VIII are generally negative or, in some cases, focally positive (Barnes 2001). The reticulin pattern is distinctive (Barnes 2001). With the exception of CD34, which may stain tumour cells, vascular markers (factor VIII, CD31) stain only the endothelial cells of the blood vessels (Barnes 2001). The tumour that most closely resembles HPC is the solitary fibrous tumour (SFT), whose cells also stain for vimentin and CD34 (Kempson et al. 2001). There is, moreover, a considerable overlap between the histological features of HPC and of SFT (Kempson et al. 2001, Veltrini et al. 2003). Some differences, however, exist and can help in the differential diagnosis. Generally, HPC shows homogeneously higher cellularity and staghorn-like vessels throughout the lesion, whereas SFT shows varying cellularity and often thick and keloid-like hyalinization (Ogawa et al. 2003). Moreover, in SFT, the tumour cells are separated by linear rows of collagen, at least focally, a feature not found usually in HPC (Kempson et al. 2001). Herringbone formations, neurofibroma- and schwannoma-like areas, and diffuse sclerosing areas may be present in SFT, but usually not in HPC (Veltrini et al. 2003). Numerous mast cells are, moreover, found in SFT and not in HPC (Veltrini et al. 2003).

In our case, the clinical differential diagnosis was made in particular with

peripheral giant cell granuloma that appears as an asymptomatic gingival red lesion with a broad-based aspect, with peripheral fibroma that appears as a firm, asymptomatic, sessile or pedunculated tumescence, with a colour similar to the surrounding tissues, and with exostosis, that is a bony hard nodule covered by intact mucosa found attached to the buccal aspect of the alveolar bone (Langlais & Miller 1992).

The prognosis of HPC is unpredictable but usually favourable (Ogawa et al. 2003, Veltrini et al. 2003). All HPCs have an innate capacity to behave in a malignant fashion (Barnes 2001), even if the potential for malignant behaviour is often difficult to predict from a histological point of view (Coffin 1997, Sikes et al. 2003).

Some authors divide the HPC into benign, borderline and malignant. In the benign type, there are up to one mitosis per 20 HPF and no anaplastic changes. Borderline tumours present an increased cellularity with compression of vascular spaces, slight anaplasia and one to four mitoses per 20 HPF (Barnes 2001). Malignant tumours show three or more mitoses per five HPF (Barnes 2001). Moreover, prominent cellularity, large tumour size, increased mitotic activity, presence of haemorrhage, necrosis, cellular anaplasia and increased cellularity may be useful for assigning a grade to the tumour (Sikes et al. 2003). These features can be supplemented by determining the proliferation index using immunohistochemical techniques. A proliferation index of 10% or greater, as measured with MIB-1, may be indicative of a more aggressive subset of this neoplasm (Kowalski & Paulino 2001). HPCs of the head and neck are believed to behave more favourably compared with their counterparts in other anatomic sites (Walike & Bailey 1971, Llorente et al. 1999, Enzinger & Weiss 2001). Recurrence rate has been reported to be as high as 57% (Barnes 2001).

Treatment is primarily surgical, although some reports advocate the use of radiation therapy in those patients with a high-grade lesion or incomplete resections (Staples et al. 1990, Kowalski & Paulino 2001).

The reported metastases vary from approximately 10–60% (Sikes et al. 2003) and they can occasionally occur up to 10 years after surgery. They are usually located in the lungs, liver and bones; lymph node metastasis is uncommon. Overall, the 10-year actuarial survival was reported to be 70% for tumours with zero to three mitoses per 10 HPF and 29% for tumours with four or more mitoses per 10 HPF (Barnes 2001).

In conclusion, HPC is an uncommon vascular tumour in which the biological behaviour is difficult to predict, and the patients need to be followed up closely for an extended period (Watanabe et al. 2001).

Acknowledgements

This work was partially supported by the National Research Council (CNR), Rome, Italy, by the Ministry of Education, University and Research (MIUR), Rome, Italy, and by AROD (Research Association for Dentistry and Dermatology), Chieti, Italy.

References

Abdel-Fattah, H. M., Adams, G. L. & Wick, M. R. (1990) Hemangiopericytoma of the maxillary sinus and skull base. *Head and Neck* **12**, 77–83.

Alabdulhadi, K., Burezq, H., Nguyen, V. H., Bernard, C. & Manoukian, J. J. (2004) Extensive tongue hemangiopericytoma in a child: modified combined modality of treatment to preserve well functioning tongue. *International Journal of Pediatric Otorhinolaryngology* **68**, 211–219.

August, L. J., Razack, M. S. & Sako, K. (1982) Hemangiopericytoma. *Journal of Surgical Oncology* **20**, 260–264.

Barnes, L. (2001) Tumors and tumor-like lesions of the soft tissues. In: Barnes, L. (ed) *Surgical Pathology of the Head and Neck*, 2nd edition, pp. 890–1048. New York: Marcel Dekker Inc.

Carew, J. F., Singh, B. & Kraus, D. H. (1999) Hemangiopericytoma of the head and neck. *Laryngoscope* **109**, 1409–14111.

Coffin, C. M. (1997) Vascular tumors. In: Coffin, C. M., Dehner, L. P. & O'Shea, P. A. (eds). *Pediatric Soft Tissue Tumors. A Clinical, Pathological and Therapeutic Approach*, 1st edition, pp. 40–79. Baltimore: Williams and Wilkins.

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

Enzinger, F. M. & Weiss, S. W. (2001) *Soft Tissue Tumors*, 4th edition, pp. 1004–1019. St. Louis: Mosby-Year Book.

Goldwasser, M. S. & Daw, J. L. (1990) Hemangiopericytoma of the palate: case report. *Journal of Oral and Maxillofacial Surgery* **48**, 221–215.

Hiraumi, H., Kitajiri, S., Hirose, T., Hosaka, N., Toki, J. & Ikehara, S. (2002) Radiosensitive hemangiopericytoma in the soft palate. *Auris Nasus Larynx* **29**, 95–97.

Kempson, R. L., Fletcher, C. D. M., Evans, H. L., Hendrickson, M. R., & Sibley, R. K. (2001) *Tumor of the Soft Tissue by Atlas of Tumor Pathology*. Bethesda: Armed Forces Institute of Pathology.

Kowalski, P. J. & Paulino, A. F. (2001) Proliferation index as a prognostic marker in hemangiopericytoma of the head and neck. *Head and Neck* **23**, 492–496.

Langlais, R. P. & Miller, C. S. (1992) *Color Atlas of Common Oral Diseases*, pp. 20–21. Philadelphia: Lea and Febiger.

Llorente, J. L., Suarez, C., Ablanedo, P., Carreno, M., Alvarez, J. C. & Rodrigo, J. P. (1999) Hemangiopericytoma of the parapharyngeal space. *Otolaryngology – Head and Neck Surgery* **120**, 531–533.

McMaster, M. J., Soule, E. H. & Ivins, J. C. (1975) Hemangiopericytoma. A clinicopathologic study and long-term follow-up of 60 patients. *Cancer* **36**, 2232–2244.

Ogawa, I., Sato, S., Kudo, Y., Miyauchi, M., Sugiyama, M., Suei, Y. & Takata, T. (2003) Solitary fibrous tumour with malignant potential arising in sublingual gland. *Pathology International* **53**, 40–45.

Omer, G., Fetin, R. & Naki, S. (1995) Mandibular hemangiopericytoma: report of a case and review of the literature. *Journal of Oral and Maxillofacial Surgery* **53**, 704–707.

Sellke, F. W., Laszewski, M. J., Robinson, R. A., Davis, R., & Rossi, N. P. (1991) Hemangiopericytoma of the sternum. *Archives of Pathology and Labortory Medicine* **115**, 242–244.

Sikes, J. W., Ghali, G. E., Veillon, D. M., & Buchbinder, D. (2003) Parotid mass in a 62-year-old man. *Journal of Oral and Maxillofacial Surgery* **61**, 1073–1077.

Singh, B., Govender, S. & Joshi, S. D. (1993) Hemangiopericytoma of the floor of the mooth. A case report. *South African Journal of Surgery* **31**, 139–141.

Staples, J. J., Robinson, R. A., Wen, B. C. & Hussey, D. H. (1990) Hemangiopericytoma-the role of radiotherapy. *International Journal of Radiation Oncology Biology Physics* **19**, 445–451.

Stout, A. P. (1942) Hemangiopericytoma: a vascular tumor featuring Zimmermann's pericytes. *Annals of Surgery* **116**, 26–33.

Veltrini, V. C., Etges, A., Magalhaes, M. H. C. G., de Araujo, N. S. & de Araujo, V. C. (2003) Solitary fibrous tumor of the oral mucosa. Morphological and immunohistochemical profile in the differential diagnosis with hemangiopericytoma. *Oral Oncology* **39**, 420–426.

Vogler, J. J., Andavolu, R. & Leban, S. G. (1990) Malignant hemangiopericytoma of the gingiva: report of a case. *Journal of Oral and Maxillofacial Surgery* **48**, 990–992.

Volpe, A. G., Sullivan, J. G. & Chong, F. K. (1991) Aggressive malignant hemangiopericytoma in the neck. *Journal of Surgical Oncology* **47**, 136–138.

Walike, J. W. & Bailey, B. J. (1971) Head and neck hemangiopericytoma. *Archives of Otolaryngology* **93**, 345–353.

Watanabe, K., Saito, A., Suzuki, M., Yamanobe, S. & Suzuki, T. (2001) True hemangiopericytoma of the nasal cavity. Immunohistochemical and electron microscopic study of 2 cases and a review of the literature on sinonasal hemangiopericytomas. *Archives of Pathology & Labortory Medicine* **125**, 686–690.

Wold, L. E., Unni, K. H., Cooper, K. L., Sim, F. H. & Dahlin, D. C. (1982) Hemangiopericytoma of bone. *American Journal of Surgical Pathology* **6**, 53–58.

Address:
Adriano Piattelli
Via F. Sciucchi 63
66100 Chieti
Italy
E-mail: apiattelli@unich.it

Clinical Relevance

Scientific rationale: HPC represents about 3% of all tumours in the head and neck. Because of the rarity of this lesion, data collection on tumour behaviour and appropriate treatment is feasible only through case reports.

Principal findings: we decided to report a case of HPC, which should be of interest to a Periodontist for its location (maxillary gingiva) and its clinical differential diagnosis with more commonly collected lesions such as exostosis, peripheral fibroma and peripheral giant cell granuloma.

Practical implications: awareness of this lesion combined with a careful histological analysis can help clinicians to avoid potential diagnostic pitfalls and to pursue the appropriate treatment.