

Case report**Hemangiopericytoma of gingiva- A Case Report**

Raj A, Ramesh G, Pathak S, Sant V

ABSTRACT: Hemangiopericytoma/solitary fibrous tumor is a very rare tumor of uncertain malignant potential. Under the World Health Organization (WHO) classification, hemangiopericytomas and solitary fibrous tumors of the soft tissues and are regarded as features of the same entity in the soft tissue fascicle. Stout and Murray described these tumors as "vascular tumors arising from Zimmerman's pericytes". Head and neck lesions represent 16-25% of all reported hemangiopericytomas, and they represent 2-3% of all soft tissue sarcomas in humans. Here we report a case of hemangiopericytoma of attached gingiva which clinically appeared as pyogenic granuloma.

Key words: Hemangiopericytoma; Vascular Tumor; Pericytes; Pyogenic Granuloma; Soft Tissue Sarcoma.

INTRODUCTION

Hemangiopericytomas (HPC) are soft tissue sarcomas that originate in pericytes, cells of mesenchymal origin that partially surround the endothelial cells of capillaries and small veins where they assist in the regulation of blood flow.¹ Origin of this tumor has been attributed to Chromosomal translocations t(12;19) and t(13;22) in lesional cells.² They are usually considered to be benign neoplasm, but has a definite malignant counterpart.

In the 2002 WHO classification,³ it is regarded as a tumor with potential low malignancy. Stout was the first to report an oral hemangiopericytoma.¹ We reported a similar case of oral hemangiopericytoma presenting itself clinically, as a pyogenic granuloma.

CASE REPORT

A male patient aged 22 yrs, reported to a private dentist with a swelling and bleeding gums in right maxillary premolar and first molar area since one year. On examination, a growth was seen in 15, 16 region, which was pinkish red in colour, roughly oval in shape, smooth surface and approximately 2cms in size. Base of the lesion was sessile and did not bleed profusely on probing. Excisional biopsy was done and the tissue was submitted for histopathological examination to the department of Oral Pathology, Rama Dental College, Hospital and Research

Centre. A provisional diagnosis of pyogenic granuloma was made.

On histopathologic examination, the H & E stained section revealed the presence of hyperplastic parakeratinized stratified squamous surface epithelium with elongated rete-ridges. The subjacent connective tissue stroma was fibrocellular with plenty of endothelial lined capillaries of varying shapes and sizes (Figure 1).

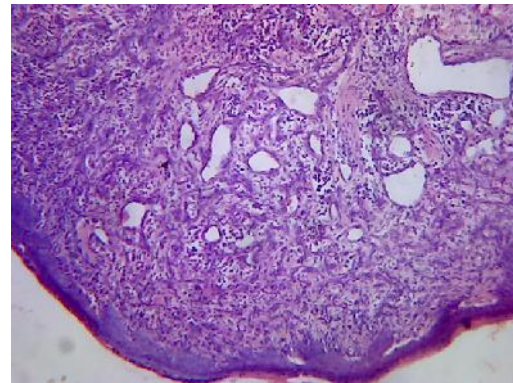


Fig 1- H&E stained section (10x magnification) shows the plenty of blood capillaries of varying sizes and shapes surrounded by sheets of spindle shaped cells.

Few capillaries showed the branching pattern simulating staghorn-like pattern with mixed hyper- and hypo-cellular areas. Few capillaries showed peripheral proliferation of spindle cells (Figure 2&3). An area with ulceration was seen showing dense infiltration of chronic inflammatory cells.

Based on these findings, a confirmatory diagnosis of hemangiopericytoma was made.

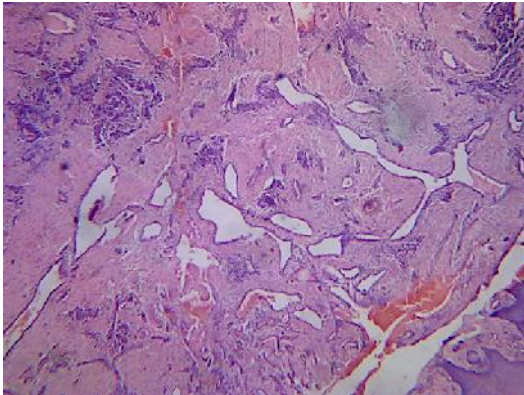


Fig 2- H&E stained section (10x magnification) shows the plenty of blood capillaries with less cellularity with branching pattern of few capillaries.

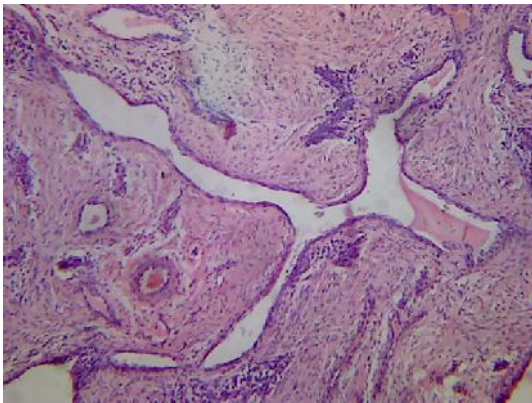


Fig 3- H&E stained section (40x magnification) shows typical staghorn pattern of the blood capillary as seen in classic HPC.

DISCUSSION

Hemangiopericytomas are soft tissue sarcomas that originate from pericytes, also known as Rouget cells or mural cells, are contractile cells that wrap around the endothelial cells of capillaries and small veins where they assist in the regulation of blood flow. These pericytes are embedded in basement membrane where they communicate with endothelial cells of the body's smallest blood vessels by means of both direct physical contact and paracrine signaling.⁶ Over the last decades, studies of blood vessels have concentrated mainly on

the endothelial cell component, especially when the first angiogenic factors were discovered. Pericytes are, however, functionally significant; when vessels lose pericytes, they become hemorrhagic and hyperdilated, which leads to conditions such as edema, diabetic retinopathy, and even embryonic lethality. Recently, pericytes have gained new attention as functional and critical contributors to tumor angiogenesis and therefore as potential new targets for antiangiogenic therapies.⁴

HPC's are seen usually around irregularly formed vascular tissue. They can occur in bone and soft tissue, muscle, liver, and the heart, mimicking Sarcomas. Fortunately, HPC's are rare. Reports have indicated that it tends to arise in the head and neck, but its occurrence in the mouth is regarded as rare. In the 2002 WHO classification,³ it is not categorized as neither benign nor malignant, but is regarded as a tumor with potential low malignancy. Yasuyuki Michi⁵ in 2013 reviewed and found that 16 cases of HPC's of the mouth have been reported worldwide of which eight were described as malignant.

In general, HPC's is seen to involve both sexes at equal rates and all age groups.⁵ The age ranges from 13 to 91 years, with a median age of 44.5 years, and a slightly elevated incidence among women (male:female=1:1.3). The most common site of origin was palate, followed by mandible lower lip. Clinical symptoms most commonly comprised a mass / swelling, ranging from 3 mm to 60 mm, with a median size of 23 mm, and associated pain was not reported in any of the case.⁶ The present case was a male patient in his early 20s.

HPC's consists of numerous vascular channels with plump endothelial nuclei and a surrounding, tightly packed proliferation of oval and spindled cells with dark nuclei and a moderate amount of cytoplasm. Areas with more spindled pericytes may show an interlacing pattern of cells but usually there is a medullary tissue pattern, sometimes with palisading of cells, reminiscent of a neural tumor. Older, less aggressive lesions tend to have less cellularity and may have a largely mucoid interstitial appearance, which can be

mistaken for myxoid lipoma or myxoid liposarcoma. Focal cartilage production may rarely be seen and such lesions must be differentiated from mesenchymal chondrosarcoma.⁷

Reticulin staining can be used to demonstrate lesional vessels lined by a single layer of endothelial cells, with the pericytes lying outside the basal lamina, although they are often individually surrounded by reticulin and collagen fibers. Lesional cells are immunoreactive for vimentin (variable intensity), factor XIIIa antigen, HLA-DR antigen and CD34.⁷

Although the 2002 WHO classification does not include clear diagnostic criteria for the grades of malignancy,⁸ characteristic of malignant hemangiopericytomas are described in the literature. These include increased cellular density and hemorrhage, necrosis, cellular atypia, nuclear polymorphism and elevated mitotic figures.⁹ Enzinger and Smith⁵ and GüerriSSI *et al.*¹⁰ reported that mitotic figures of 4 and 4/10 HPF, respectively, were associated with malignant lesions. Batsakis *et al.*⁹ found that mitotic figures 1/10 HPF and 1/20 HPF, in addition to mild or moderate cellular atypia, respectively, were consistent with malignant tumors.

HPC's resemble to many spindle cell tumors. Hence other spindle cell lesions to be considered for differential diagnosis are fibrous histiocytoma, MFH, synovial sarcoma, other stromal sarcomas, juxtaglomerular tumor, vascular leiomyoma, and juvenile hemangioma.

Histopathologically, the so-called 'stag-horn' sign, formed by proliferation of fusiform to roundish undifferentiated tumor cells in dendritic branches around the capillary vessels, was formerly regarded¹⁰ as useful in the diagnosis of HPC's. However, because this finding is also present in many other soft-tissue tumors, it is no longer considered a distinguishing characteristic of HPC's. In order to distinguish HPC's, from other solitary fibrous tumors, diagnosis must be based on results of HPC's.¹¹

Solitary fibrous tumors (SFTs) have a broad histological spectrum with appearances often varying from field-to-field within one tumor, thus contributing to diagnostic difficulties. In the mid-1990s, the diagnosis of HPC's was called into question¹², and in 2002, the WHO acknowledged that the majority of tumors formerly diagnosed as HPC could be reclassified as any number of other soft tissue tumors including SFTs. Many pathologists now believe that the diagnosis of HPC should be used only for truly pericytic lesions, such as the sinonasal HPC. Chan¹³ suggested several diagnostic criteria for SFT, namely:

1. Circumscription
2. Alternating hypercellular foci and hypocellular sclerotic foci
3. Short spindly or ovoid cells with scanty and poorly defined cytoplasm
4. Few mitotic figures (<4/10 HPF)
5. Intimate intertwining of thin or thick collagen fibrils with spindle cells
6. CD34 positivity of spindled cells

SFT is a mesenchymal neoplasm of fibroblastic and not mesothelial origin. The non-pleural tumors that resembled HPC as described by Stout are mostly believed to represent extra-pleural SFTs and many have abandoned HPC as a diagnostic term in favor of the term SFT. There is a residual group of tumors that currently retain the diagnosis of HPC and these include sinonasal HPC, which demonstrates cells with true pericytic properties. This group of tumors of pericytic origin in future will likely be reclassified as myopericytomas.

Solitary fibrous tumors showed cellularity and collagenization varying from area to area, focal perivascular hyalinization, scattered giant nuclei cells and abundant mast cells throughout the tumor. The HPC's exhibits thin-walled and dilated vessels lined with flat endothelial cells, identified by "staghorn appearance".¹⁴ Tumoral cells of solitary fibrous tumor exhibit immunohistochemical positivity for CD34, as well as endothelial cells.

The hemangiopericytoma is positive only in endothelial cells. In solitary fibrous tumor,

alpha-smooth muscle actin, h-caldesmon and laminin stained the wall vessels. In HPC's, on the other hand, the wall vessels were positive only for laminin, which staining was also observed in perivascular tumoral cells. These morphological and immunohistochemical differences infer these lesions constitute distinct entities.¹⁴

CONCLUSION: Vascular anomalies are congenital frequently involve the head, neck, and oral cavity. Subdivided into vascular tumors (hemangiomas) and vascular malformations, vascular anomalies remain poorly understood.¹⁵ Any pyogenic granuloma-like lesion should be palpated carefully, and if a nodule is present, surgical excision rather than cryotherapy or laser ablation should be considered.¹⁶ Because of the rarity and unpredictable biological behavior¹⁷ of these tumors, long-term follow-up is necessary even after radical resection because recurrence or development of metastasis may be delayed by many years.

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REFERENCES

1. Stout AP, Murray MR. Hemangiopericytoma: vascular tumor featuring Zimmermann's pericytes. *Ann Surg* 1942; 116:26
2. Rajendran R. Benign and Malignant Tumors of the Oral Cavity. In Rajendran R, Sivapathasundharam B. Shafer's textbook of oral pathology. 6thed. Reed Elsevier India Private Ltd 2006;162-163.
3. Goldwasser MS, Daw JL. Hemangiopericytoma of the palate: case report. *J Oral Maxillofac Surg*. 1990;48 2:211–215.
4. Bergers G and Song S. The role of pericytes in blood-vessel formation and maintenance. *Neuro Oncol*. Oct 2005; 7(4): 452–464.
5. Michi Y, Suzuki M, Kurohara K, Harada K. A case of hemangiopericytoma of the soft palate with articulate disorder and dysphagia *Int J Oral Sci*. Jun 2013; 5(2): 111–114.
6. Enzinger FM, Smith BH. Hemangiopericytoma. An analysis of 106 cases. *Hum Pathol*. 1976;7 1:61-82.
7. Saify F, Sharma N. Hemangiopericytoma of Buccal Mucosa: Report of a Rare Case with Review of Literature. *Indian J Dent Adv* 2012; 4(4): 1011-1015
8. Fletcher CD, Unni K, Mertens F. World health organization. Classification of tumours, pathology and genetics, tumours of soft tissue and bone. Lyon; IARC Press; 2002. p. 432.
9. Batsakis JG, Rice DH. The Pathology of head and neck tumors: vasoformative tumors, part 9B. *Head Neck Surg*. 1981;3 4:326–339.
10. GüerriSSI JO, Miranda MG, Olivier R. Giant hemangiopericytoma of mandible: a propos of a case: a variant of the surgical technique for protection of the articular fosse. *J Craniofac Surg*. 2006;17 3:523–527.
11. Stout AP. Hemangiopericytoma: a study of twenty five new cases. *Cancer*. 1949;2 6:1027–1035.
12. Regan EMO, Vanguri V, Allen CM, Eversole LR, Wright JM, Woo SB. Solitary Fibrous Tumor of the Oral Cavity: Clinicopathologic and Immunohistochemical Study of 21 Cases. *Head Neck Pathol*. Jun 2009; 3(2): 106–115.
13. Chan JKC. Solitary fibrous tumour everywhere and a diagnosis in vogue. *Histopathology*. 1997;31(6):568–576.
14. Veltrini VC, Etges Magalhães MH, de Araújo NS, de Araújo VC.A. Solitary fibrous tumor of the oral mucosa--morphological and immunohistochemical profile in the differential diagnosis with hemangiopericytoma. *Oral Oncol*. 2003 Jun;39(4):420-426.

15. Buckmiller LM, Richter GT, Suen JY. Diagnosis and management of hemangiomas and vascular malformations of the head and neck. *Oral Dis.* 2010 Jul;16(5):405-418.
16. Hung CH, Kuo HW, Chiu YK, Huang PH. Intravascular pyogenic granuloma arising in an acquired arteriovenous malformation: report of a case and review of the literature. *Dermatol Surg.* 2004 Jul;30(7):1050-1053.
17. Angiero F, Signore A, Benedicenti S. Hemangiopericytoma/Solitary fibrous tumor of the oral cavity. *Anticancer Res.* 2011 Feb;31(2):719-723.

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