

CASE REPORT

MALIGNANT HEMANGIOPERICYTOMA OF PALATE- A CASE REPORT

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ABSTRACT:

Hemangiopericytoma (HPC) is a rare tumor of uncertain malignant potential. Stout and Murray described HPC as “vascular tumor arising from Zimmerman’s pericyte” in 1942. The World Health Organization (WHO) reclassified HPC as a fibroblastic/ myofibroblastic tumor, after further characterization. HPC is found mostly wherever there is increased vascularity seen. The incidence of the tumor in head and neck area is only 15%, mostly seen in adults. We report here a case of HPC of a 26-year-old male, who presented to our department with a non-tender swelling in maxillary posterior region and the mass was well-circumscribed, pedunculated and soft on palpation. Overlying mucosa appears red and erythematous with normal surrounding mucosa with irregular margins. The tumor was completely removed with wide surgical resection. The histopathological staining supported the diagnosis of HPC.

Key words: Hemangiopericytoma, pericytes.

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CASE REPORT

A 26 yr old male reported with the complaint of swelling at the right & left side of posterior maxilla, since 3 month. On examination, palatal aspect of 27 A solitary over growth seen on left back tooth region measuring approx 3 x 2cm in its longest dimension, extending mesiodistally from mesial aspect of 25 till distal aspect of 27 covering the mesial half of occlusal surface and buccopalatally involving the buccal vestibule extending palatally 1cm away from the midline. Overlying mucosa appears red and erythematous with normal surrounding mucosa with irregular margins. Another a well defined solitary overgrowth seen on the palatal aspect of 17, 18 extending from marginal gingiva measuring approx 1.5 x 1 cm in its longest dimension with normal overlying mucosa surrounded by an erythematous zone (**Figure 1**). Excisional biopsy was done and the tissue was submitted for histopathological examination to the department of Oral Pathology, Rungta College of Dental Science 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 broad rete-ridges. The subjacent connective tissue stroma was fibrocellular with plenty of endothelial lined capillaries of varying shapes and sizes. 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**). An area with ulceration was seen showing dense infiltration of chronic inflammatory cells.



Figure 1: An overgrowth was present on palatal aspect of 27 extending till buccal vestibular region of 27.

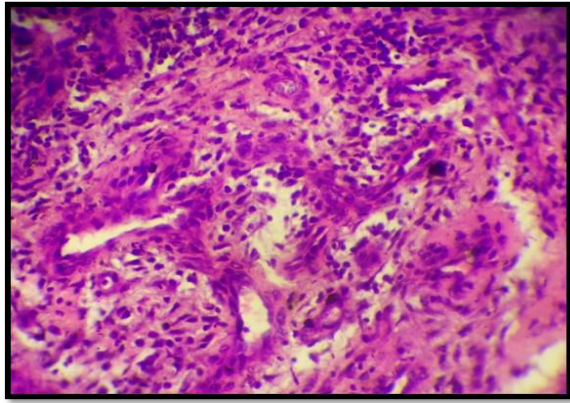


Figure 2: H&E stained section (10x magnification) shows the plenty of blood capillaries ,surrounded by proliferated pericytes

DISCUSSION

A hemangiopericytoma is a rare, soft-tissue tumor of vascular origin derived from a pericyte of Zimmerman, which is a modified smooth muscle cell that surrounds the small blood vessels. This type of tumor was first described by Stout and Murray in 1942. In 1923, Zimmerman first described the **pericyte** as “a smooth muscle-related cell with contractile powers, although lacking myofibrils, and long processes that wrap around capillaries to change the lumen caliber.”¹ 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.² 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. 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 classification³, it is not categorized as neither benign nor malignant, but is regarded as a tumor with potential low malignancy. Similar to the case presented here in, HPC usually presents as a slow growing painless mass associated with only local symptoms. Yasuyuki Michi 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 1949, Stout expanded on his previous work by better delineating the histological details of 25 cases of

HP submitted to him from medical centers around the country. Two of these originated in the head and neck; the first reported case was of nasal HP, another was in the tongue base. Microscopically, the tumors consisted of tightly packed cells around endothelially lined vascular channels. In general, HPC's is seen to involve both sexes at equal rates and all age groups. 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.³ The present case was a male patient in his early 20s. The etiology is unknown, although HP has been linked to trauma, prolonged steroid use, and hormonal imbalance. 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.⁴ Although the 2002 WHO classification does not include clear diagnostic criteria for the grades of malignancy,⁵ 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.⁶ Enzinger and Smith and Güerrissi *et al.* reported that mitotic figures of 4 and $\geq 4/10$ HPF, respectively, were associated with malignant lesions. Batsakis *et al.*⁷ found that mitotic figures $\geq 1/10$ HPF and $\geq 1/20$ HPF, in addition to mild or moderate cellular atypia, respectively, were consistent with malignant tumors. **According to Barnes**, HPC's have three types regarding the malignant potential:

- Benign type, in which there are up to one mitosis per 20 HPF (high power fields) and no anaplastic changes.
- Borderline type, which present an increased cellularity with compression of vascular spaces, slight anaplasia and one to four mitoses per 20 HPF.
- Malignant type, which show three or more mitoses per five HPF.⁹

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 regarded 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.⁷ Diagnosis of HPC should be used only for truly pericytic lesions, such as the sinonasal HPC. Chan 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". Tumoral cells of solitary fibrous tumor exhibit immunohistochemical positivity for CD34, as well as endothelial cells.

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. 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. 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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