Journal of Advanced Medical and Dental Sciences Research

@Society of Scientific Research and Studies

Journal home page: www.jamdsr.com doi: 10.21276/jamdsr UGC approved journal no. 63854

(e) ISSN Online: 2321-9599; (p) ISSN Print: 2348-6805

Case Report

Hemangiopericytoma of Buccal Mucosa- A Case Report

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ABSTRACT:
A Hemangio-pericytoma (HPC) is a soft tissue sarcoma that originates in the pericytes in the walls of capillaries. It was described by Stout and Murray in 1942. It appears as a painless mass and is observed mainly in the sixth and seventh decades of life. It is an unknown occurrence in head and neck region. We report a 32-year-old female who presented with a hemangiopericytoma in the left buccal mucosa.
Key words: Solitary fibrous tumor, Buccal mucosa, Cheek, vascular, pericytoma.

Received: 4 February, 2019 Revised: 25 February, 2019 Accepted: 26 February, 2019

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This article may be cited as: Bhagat N, Mukherjee T, Sharma R, Kumar S, Gupta P, Gupta R, Sharma A. Hemangiopericytoma of Buccal Mucosa- A Case Report. J Adv Med Dent Scie Res 2019;7(3):21-23.

INTRODUCTION

Hemangiopericytoma is a solitary fibrous tumor which is a very rare tumor of unknown malignant potential. In 1942, Stout and Murray described these tumors as "vascular tumors arising from Zimmerman's pericytes" They were originally described by Roughet in 1873 as pericapillary ameboid cells. Hemangiopericytoma can be found at any site of the body where pericytes are present and it mainly affects adults .This tumor rarely occurs in the oral cavity as the pericytes line the epithelial cells in the capillary walls.[1,2] We report a 32-year-old female who presented with a hemangiopericytoma in the left buccal mucosa.

CASE REPORT

A 32 year old female presented with a gradually enlarging swelling in her left buccal mucosa region since 6 months. The swelling was asymptomatic in nature, with no complains of pain or tenderness. The lesion was well circumscribed and pedunculated and the overlying mucosa presented with dilated vascular channels. The mass was rubbery in consistency with no signs of lymphadenopathy (Figure 1).The patient noticed the swelling about 7 months back when the swelling was very small in diameter of about 0.5 cm X 0.5 cm in size. The patient was undertaking homeopathic treatment for 4 months when the patient noticed the swelling to be increasing in size, the patient opted to report to our institution (School of Dental Sciences & Research, Sharda University, Greater Noida) with the lesion to be at its present size of about 1.5 cm X 1.5 cm. The adjacent structures appeared to be normal. She had no relevant medical history or any history of trauma. After a through clinical workup and hematological investigations, surgery was planned under local anesthesia for surgical excision with wide surgical margins of 0.5 cm (Figure 2). A written consent from the patient was taken for both surgical procedure and for use of the photographs for scientific publication.

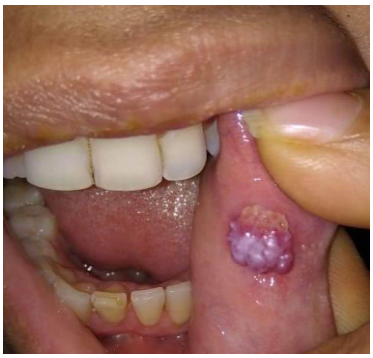

Figure 1: Intra-oral photograph

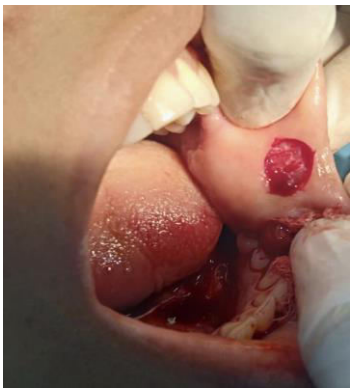

Figure 2: Surgical excision of lesion

HISTOPATHOLOGICAL EXAMINATION

H& E stained biopsy section showed overlying stratified squamous epithelium. Underlying connective tissue stroma showed fibro cellular and areas of numerous cellular proliferation surrounded by hyalinised stroma. These pathognomic areas contain vessels, furrowing capillaries with endothelial cells and surrounded by huge proliferation of pericytes (stag horn pattern).Surrounded pericytes are plump to spindle shaped along with few mitotic activities suggestive of "Benign Hemangio-pericytoma" (Figure 3).

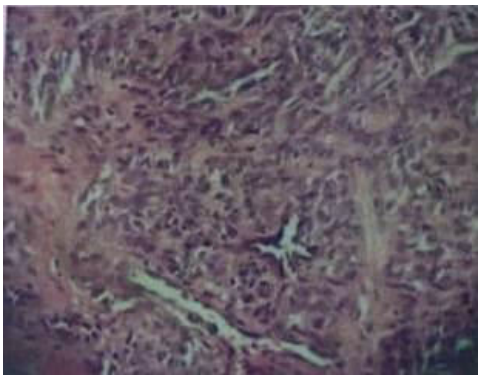

Figure 3: Histopathologic photograph

DISCUSSION

Hemangio-pericytoma(HPC) are uncommon vascular neoplasm's derived from capillary pericytes. Pericytes were first described in 1923 by Zimmerman as smooth muscle related cells that exhibit contractile function[1]. It can appear at any site where blood vessels are present and these tumors are commonly observed in the pelvis and limbs, but not frequently in the oral cavity. HPC's of head and neck are usually seen in the scalp, face, neck, oral cavity, nasal cavity, orbit and the paranasal sinuses.[2] It appears as a painless mass and is observed mainly in the sixth and seventh decades of life. It has no sex predilection. Multiple lesions are uncommon. It mainly occurs in adults and very rare in children. The tumor is closely related to the glomus tumor, which is also derived from pericytes. In cases of HPC no clear sign of its vascular nature is evident except the presence of few dilated vessels. These tumors have high tendency of malignancy and shows encapsulation despite its malignant behavior.[2]

Histopathological investigations can provide a definitive diagnosis with characteristic vascular features of anatomizing vessels, increased cellularity and prominent mitotic behavior. The tumor cells have oval nuclei and indistinct cytoplasm. However, recent immunohistochemical evidence suggests that conceptually this tumor is not derived from the pericytes because it does not express actin or myofibroblastic markers. It is likely that the neoplasm is an undifferentiated fibroblastic cell. It has been suggested that many tumors that were diagnosed previously as HPC histologically may represent other soft tissue tumors that share similar features. There is considerable histological overlap between myofibroma, solitary fibrous tumor, and chondrosarcoma of mesenchymal origin that's why for definitive diagnosis histopathological examination is of utmost importance, as HPC is a diagnosis of exclusion.[2]

The primary treatment of hemangiopericytoma is surgical excision. Chemotherapy and radiotherapy radiotherapy and chemotherapy are not as effective. The role of radiotherapy has been questioned as these tumors are generally radio resistant.[3]The time of recurrence seen in previous literature is of approximately 5 years after surgery, and 24 % of recurrences occur within one year[3]. Among the cases surveyed in the literature (27 cases), 21 patients (77.8 %) underwent surgical resection, 3 (11.1 %) underwent a combination of surgical resection, chemotherapy and radiotherapy, 2 (7.4 %) received radiotherapy alone and only one (3.7 %) received a combination of chemotherapy and radiotherapy.[4] All the benign cases were surgically resected. In head and neck region, cervical lymphadenectomy is reserved for those instances where palpable lymphadenopathy is coexistent.

In the present case, the tumor was excised as a single mass and was found to be benign.[5,6] Radiotherapy was not advised as there was no lymphadenopathy. The

patient has been kept under observation and regular follow-up, and remains free of recurrence 24 months after surgery. As a matter of caution, the tumor location and clinical characteristics[5, 6] of each hemangiopericytoma need to be followed up.

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Source of support: Nil **Conflict of interest:** None declared