Type of the manuscript: Case Report

Hemangiopericytoma of the Gingiva – A Case Report

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Abstract

Hemangiopericytoma (HPC) is a very rare tumor with uncertain malignant potential. It is a soft tissue tumor which originates from the pericytes in the walls of the blood capillaries. Both benign and malignant varieties are present with the tendency to metastasize. It was first described by Stout and Murray in 1942. The incidence of occurrence in oral cavity is very rare. We report a case of 57 year old male, who presented with an exophytic overgrowth from the lingual aspect of the maxillary left gingiva which extended into the palatal area. The tumor was completely removed with wide surgical excision. The histopathological study of the tumor suggested the HPC.

Key words: *pericytes, hemangiopericytoma, staghorn pattern.*

Introduction

Hemangiopericytoma (HPC) is a soft tissue tumor arising from pericytes, which are modified smooth muscle cells in the periphery of blood vessels. The pericytes are located outside the reticulin sheath of the endothelium.[1,2] Pericytes are the small cells having oval or spindle shaped morphology lining the capillaries.[3]

Chromosomal translocations t(12;19) and t (13;22) in the lesional cells have been related to the origin of this tumor.[4] In the WHO classification 2002, it was considered as a tumor with potential low malignancy.[5] The tumor is more common in lower extremities, pelvis and retroperitonium while very few cases (15-20%) occur in the head and neck region.[6] HPC do not show any gender predilection with most of the cases occurring in the 4th to 6th decades of life.[7] They are very uncommon in children and only accounts for about 10% of the cases.[8] We report a case of HPC in a 57 year old male along with the review of literature.

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Case Report

A 57 year old male reported with the chief complaint of bleeding gums and enlargement of gingiva in the left anterior maxillary region since 1 month. Intra oral examination revealed the presence of exophytic gingival overgrowth seen on the lingual aspect in relation to first and second premolars. The growth was solitary, measuring about 4x3.5cm in size. The overlying surface of the lesion was irregular and covered with food debris. The color of the lesion was normal in comparison to the adjacent mucosa. Few areas of the overgrowth showed indentations of the occlusal surfaces of lower teeth (Figure 1).

Blood examination revealed increase in the erythrocyte sedimentation rate to 132mm. The chest X-ray was done to rule out any metastasis, which was normal (Figure2). The CT scan examination revealed the presence of 32x22mm heterogenous well defined enhancing lesion in the left side of the oral cavity. The lesion was abutting alveolar margin of maxilla without obvious erosion or breech (Figure 3). Surgical excision of the tumor was done under sedation after getting the informed consent of the patient. The lesion was excised completely and the entire specimen was sent for histopathological diagnosis along with the differential diagnosis of peripheral giant cell granuloma and peripheral ossifying fibroma.The differential diagnosis was based on the fact that it might be a reactive lesion on the gingiva. For almost all the peripheral lesions gingiva is the most favorable site for occurrence. Reactive lesions are common tumor like proliferations within the oral cavity. They exhibit few clinical differences but their features in most occasions are very similar to the tumors. This can sometime makes it difficult for giving the differential diagnosis. According to our knowledge peripheral giant cell granuloma and peripheral ossifying fibroma are the most common differential diagnosis for peripheral lesions.

On histopathological examination, the hematoxylin and eosin stained section showed the presence of densely packed lesional cells which were round to ovoid in shape with hyperchromatic nuclei. Presence of numerous blood vessels with flat endothelial cells were seen, most of which were exhibiting typical staghorn pattern (Figure 4). There was minimal amount of any cytological atypia and mitotic activity. The final diagnosis of HPC (low grade) was done. The patient was under follow up for the next six months without any recurrence.

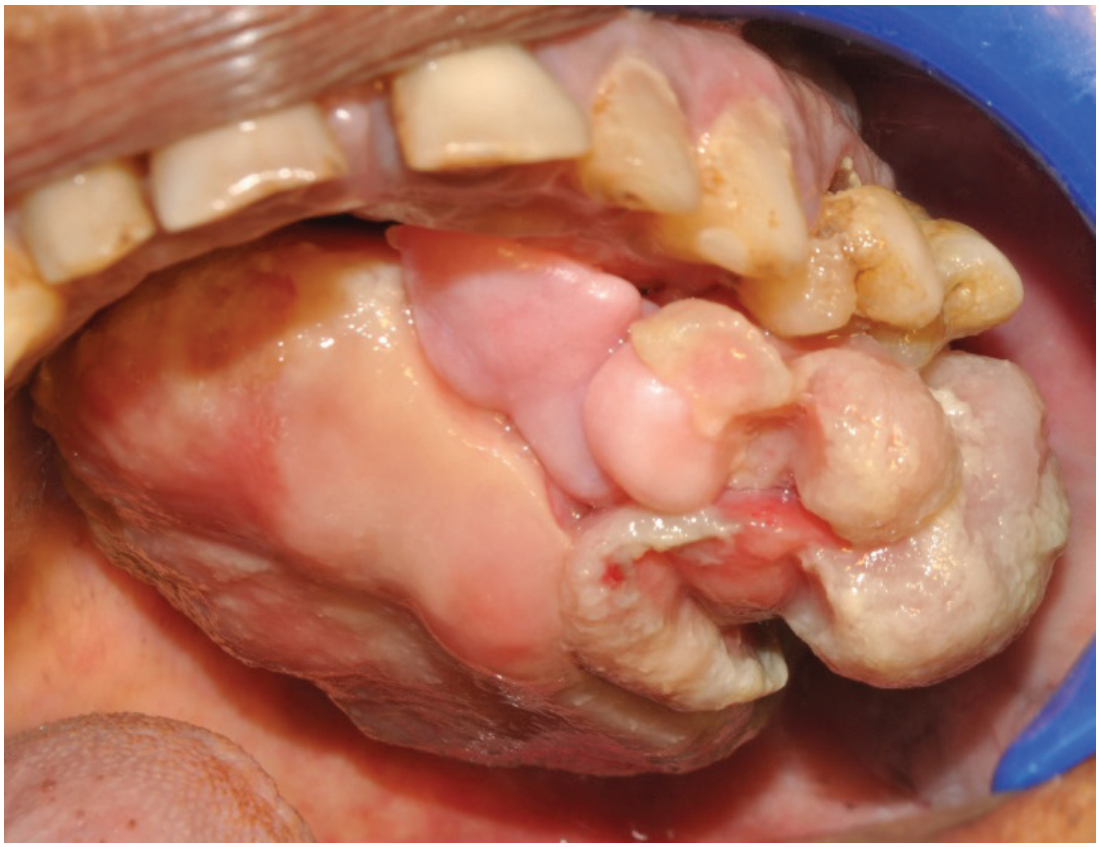

Figure 1: Exophytic gingival overgrowth

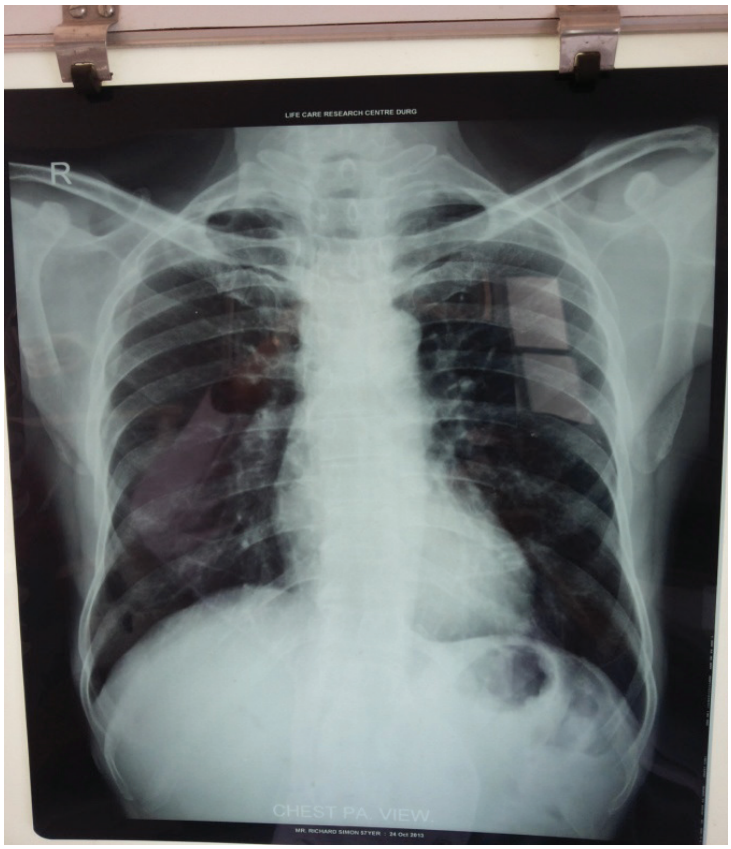

Figure 2: Chest X-ray showing no signs of metastasis

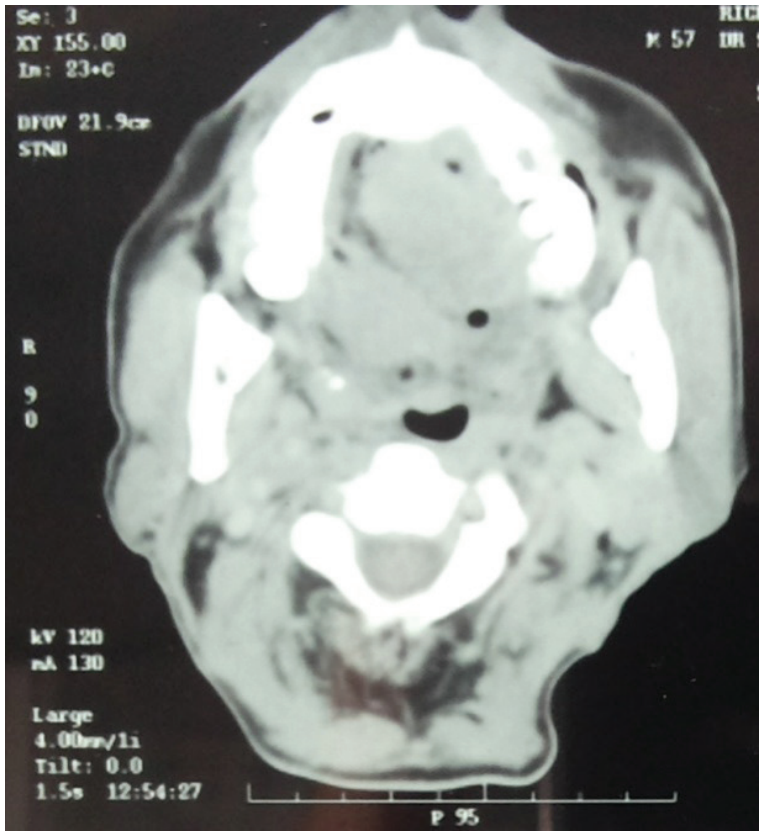

Figure 3: CT image showing well defined enhancing lesion in the left side of the oral cavity

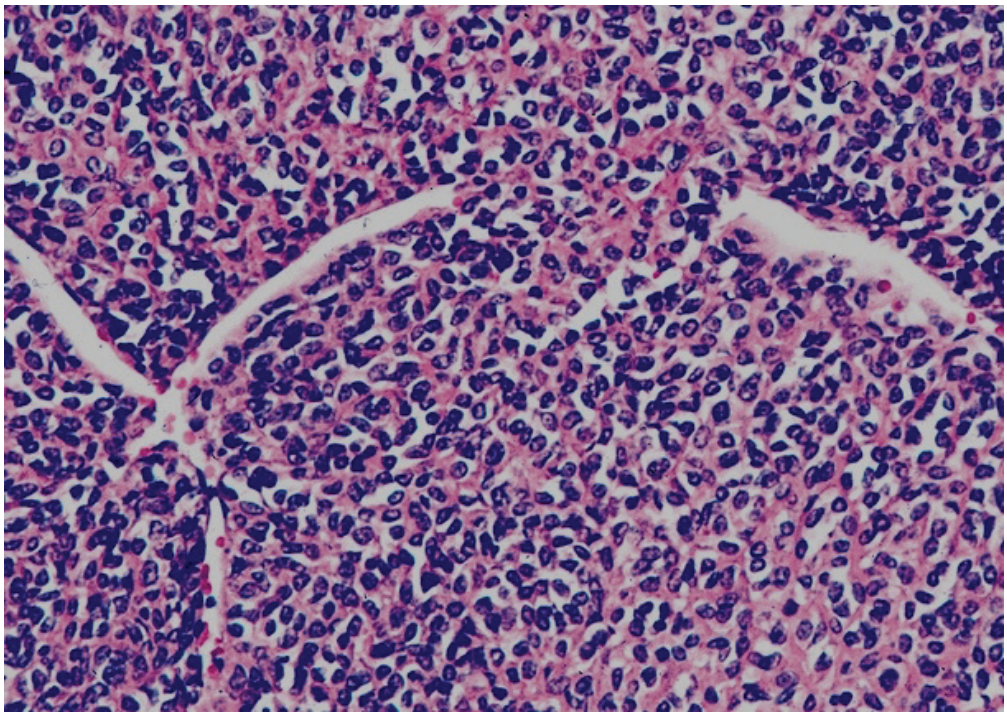

Figure 4: Characteristic staghorn pattern of blood capillaries

Discussion

Hemangiopericytoma is a soft tissue sarcoma first described by Stout and Murray in 1942. The tumor originates from the pericytes, also known as Rouget cells or Mural cells. The pericytes are the contractile cells which surround the endothelial cells of blood vessels and help in the regulation of blood flow.[9] HPC accounts for about 3-5% of all soft tissue sarcomas and about 1% of all vascular tumors. The most common sites in the head and neck region include orbit, parotid gland, nasal cavity and oral cavity. In the oral cavity, most common sites include tongue, maxilla, lips, gingiva and buccal mucosa.[1,10,11]

HPC involves both sexes with wide age range of 13 to 91 years.[12] Clinically the most common symptom comprised of swelling which in most of the cases is asymptomatic.[9]This finding was similar to our case where the patient had no complaints of any pain.The clear cut diagnostic criteria to put HPC into the category of malignancy has not been mentioned.[13] Although characteristic malignant features of HPC have been reported in the literature, which includes; necrosis, cellular atypia, nuclear pleomorphism, increased cellular density and mitotic figures.[14]The histologicaldifferential diagnosis of HPC does include fibrous histiocytoma, synovial sarcoma, vascular leiomyoma and juvenile hemangiomas as all of them also show spindle cell population.

The histological features are classified as low, intermediate and high grade, depending on the cellularity, cellular pleomorphism and mitosis. The tumor cells are ovoid or round to spindle shaped with ill-defined cell outlines. The blood vessels will be lined by endothelial cells and showing staghorn pattern.The older lesions tend to appear as less cellular with large Mucoid interstitial appearance. This may be sometimes confused with myxoid variety of lipoma or liposarcoma. Sometimes it may produce focal area of cartilage confusing it with chondrosarcoma.[1,15] In the present case similar histopathological findings were observed. The tumor showed presence of round to ovoid shaped cells with minimal amount of cellular atypia and mitotic figures. The blood vessels were arranged in a typical staghorn pattern. Overall features were suggestive of low grade HPC.

The choice of treatment for HPC is wide surgical excision. Chemotherapy and immunotherapy may be considered in case of malignant lesions. The benefit of radiation therapy is considered to be doubtful because HPC is radio resistant tumor. It may be useful in aggressive type of HPC.[1,15,16]

Conclusion

HPC is an uncommon vascular tumor with difficulty in diagnosis solely from the histopathological features. Any lesion with greater vascularity should be palpated carefully. Surgical excision of the lesion should be considered as the treatment of choice rather than the cryotherapy and laser ablation. All the vascular tumors should be kept on long term follow up to rule out the recurrence and metastasis.

Conflicts of Interest: Nil

Ethical Clearance: Since it was a case report, no ethical clearance was needed.

Source of Funding: Self

References

1. Maresi E, Tortorici S, Campione M, Buzzanca ML, Burruano F, Mastrangelo F, et al. Hemangiopericytoma of the oral cavity after a ten-year follow up. Ann Clin Lab Sci. 2007;37:274-79.
2. Kitahata Y, Yokoyama S, Takifuji K, Hotta T, Matsuda K, Tominaga T, et al. Hemangiopericytoma in the sacrococcygeal space: A case report. J Med Case Rep. 2010;4:8.
3. Tsirevelou P, Chlopsidis P, Zourou I, Valagiannis D, Skoulakis C. Hemangiopericytoma of the neck. Head Face Med. 2010;6:23.
4. Rajendran R. Benign and Malignant Tumors of the Oral cavity. In Rajendran R, Sivapathasundharam B. Shafer's textbook of oral pathology. 6th ed. Reed Elsevier India Private Ltd. 2006:162-63.
5. Goldwasser MS, Daw JL. Hemangiopericytoma of the palate: case report. J Oral Maxillofac Surg. 1990;48(2):211-15.
6. Thiele OC, Freier K, Flechtenmacher C, Rohde S, Hofele C, Muhling J, et al. Hemangiopericytoma of the mandible. J Craniomaxillofac Surg. 2010;38:597-600.
7. Sinha A, Rawson K, Singh G. A rare case of Hemangiopericytoma in the maxilla of a 4 year old child. J Indian Acad Oral Med Radiol. 2010;22:S64-66.
8. Marec-Berard P. Malignant Hemangiopericytoma. Orphanet Encyclopedia. 2004;4:1-4.
9. Enzinger FM, Smith BH. Hemangiopericytoma. An analysis of 106 cases. Hum Pathol. 1976;71:61-82.
10. Fareed MM, Al Amro AS, Akasha R, Al Assiry M, Tonio M, et al. Parapharyngeal space Hemangiopericytoma treated with surgery and post-operative radiation – A case report. Head Neck Oncol. 2012;4:10.
11. Mounayer C, Benndorf G, Bisdorff A, Wassef M, Enjolras O. Facial infantile Hemangiopericytoma resembling an arteriovenous malformation. J Neuroradiol. 2004;31:227-30.
12. 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. 2013;5(2):111-14.
13. Fletcher CD, Unni K, Mertens F. World Health Organization. Classification of tumors, pathology and genetics, tumors of soft tissue and bone. Lyon; IARC Press;2002.p.432.
14. Batsakis JG, Rice DH. The pathology of head and neck tumors: vasoformative tumors, part 9B. Head Neck Surg. 1981;34:326-39.
15. Bhutia O, Roychoudhury A. Hemangiopericytoma of the mandible. J Oral Maxillofac Pathol. 2008;12:26-28.
16. Anand R, Gupta S. Hemangiopericytoma of maxilla in a pediatric patient: A case report. J Dent Child (Chic). 2010;77:180-82.