HEMANGIOPERICYTOMA OF PALATE; A RARE CASE REPORT

Dr. Sushmita Batra¹, Dr. Pramod Krishna B², Dr. Rajdeep Singh³, Dr. Amy E. Thomas⁴ and Dr. Surabhi Singhai⁵

1. Postgraduate resident Department of oral and maxillofacial surgery Chhattisgarh dental college and research institute, Rajnandgaon, C.G.
2. Professor Department of oral and maxillofacial surgery Subbaiah institute of dental sciences, Shivamogga, Karnataka.
3. Professor Department of oral and maxillofacial surgery Chhattisgarh dental college and research institute, Rajnandgaon, C.G.
4. Assistant Professor Department of oral and maxillofacial surgery Chhattisgarh dental college and research institute, Rajnandgaon, C.G.
5. Postgraduate resident Department of oral and maxillofacial surgery Chhattisgarh dental college and research institute, Rajnandgaon, C.G.

Manuscript Info

Abstract

The Hemangiopericytoma is a rare vascular tumour originating from the pericytes, which are found on the external surface of the capillaries and are thought to act as a sphincter that controls blood flow. It has a high propensity for recurrence and metastasis. The tumor usually develops as a slowly enlarging painless mass. It typically occurs in the soft tissues of the extremities and trunk, and is rarely seen in the oral cavity. 15–16% are seen in the head and neck region. The most common sites of involvement are scalp, face, neck, nasal cavities and paranasal sinuses. Involvement of the palate is extremely rare and so here we are presenting a case of hemangiopericytoma of palate, its diagnosis, management, and a brief review of literature.

Copy Right, IJAR, 2021, All rights reserved.

Introduction:

Hemangiopericytoma is an uncommon soft tissue tumor of mesenchymal origin. It accounts for approximately 1% of all vascular neoplasms [¹]. Stout and Murray were the first to name this tumor as hemangiopericytoma [²]. The tumor is believed to arise from the pericytes of capillaries and can grow anywhere in the human body where capillaries exist. However, it is predominantly seen in the lower extremities, pelvic and retroperitoneal region. The chances of occurrence of this tumor are rare in head and neck region accounting only 15-16 % of all hemangiopericytomas [³].

This report presents an unusual case of hemangiopericytoma of hard palate of a 57-year-old male patient with a chief complaint of an overgrowth in the gum region for 15-20 days accompanied by pain. Radiographic and histopathological findings were suggestive of hemangiopericytoma of hard palate.
Case Report
A 57-year-old male patient came to the department of oral and maxillofacial surgery at our dental college with the chief complain of overgrowth and pain in the gums of upper left back teeth region in the past 3-4 months. He also complained of bad breath since that time. Patient also faced difficulty in chewing food and difficulty in speech. Gradual increase in the size of overgrowth led him to consult a doctor. There was no significant history of any systemic disease and gave no positive history of any previous trauma or infection in the same or other region. No abnormalities were found on the extraoral examination. The clinical soft tissue examination revealed a single, irregular exophytic overgrowth measuring approximately 5*6 cm in size covering almost the palatal area in relation to 24,25&26. The lesion was pedunculated and firm in consistency (Figure 1). There was slight tenderness on palpation and showed no bleeding on provocation. The patient was subjected to radiological examination for further evaluation of underlying cause. CT scan revealed a well-defined enhancing lesion along the soft palate and deviation of nasal septum towards right along with bilateral maxillary & right ethmoid sinusitis. Other findings like chest radiography, complete blood count and biochemical investigations were unremarkable.

Excisional biopsy of the lesion was planned under general anesthesia through palatal approach. Under general anaesthesia with nasotracheal intubation, the tumour was completely excised together along with resection of 2mm normal margins through a palatal approach (figure 2-A, B). Extraction of 24,25,26 & 27was also performed (figure 2-C). The excised specimen was slightly pinkish in colour and measured approximately 5*6 cm in size. Antibiotics and analgesics were prescribed postoperatively. Excised specimen was sent for histopathological examination which showed a circumscribed nodule in which densely packed cells were arranged in a haphazard pattern, surrounding thin-walled, endothelium-lined vascular channels. The vascular spaces ranged in size from small capillaries to large sinusoidal spaces. A “staghorn” vascular configuration was prevalent (figure 2- D).

![Figure 1: Exophytic overgrowth adjacent to the left maxillary 1st premolar, 2nd premolar and 1st molar: pedunculated on a broad-based stalk, 6*5 cm in size and covered with healthy mucosa.](image1)

![Figure 2: Lesion excised in toto measuring approximately 5*6 cm in size, C, extracted 24,25,26,27 & D, histopathological evaluation showing “staghorn pattern”](image2)
Patient was routinely followed up for 2-3 years but showed no recurrence. Given the absence of necrosis, the cytological homogeneity and the low mitotic index, the final diagnosis was hemangiopericytoma.

**Discussion:**

Hemangiopericytoma (HPC) was first described by Stout and Murray in 1942 as a distinctive soft tissue neoplasm, presumably of pericytic origin. Pericytes were described as smooth muscle-related cells that exhibit contractile function even though lacking myofibrils by Zimmerman in 1923\(^3\). HPC is an uncommon mesenchymal tumor, accounting for 1% of all blood vessel-related neoplasms and approximately 3% of all soft tissue sarcomas. It can occur in all age groups but more often occurring in the 5th and 6th decades of life. Only 10% of HPC occur in children. There is no sex predilection\(^4,5\).

Hemangiopericytoma commonly presents as a slowly enlarging mass which is usually painless and asymptomatic and is not noticed until it interferes with the normal function, or it is in an area where it is promptly detectable by the patient.

Pain usually occurs only in large lesions that are locally invasive or confined in unyielding spaces such as the paranasal sinuses \(^7\). Although trauma, prolonged steroid use and hormonal imbalance have been shown to be associated with HPC, but its exact etiology remains unascertained\(^6\).

A thorough histological examination of the surgically excised specimen should be performed because a simple biopsy is frequently insufficient to make the diagnosis\(^2\).

Hemangiopericytoma has a wide variation in clinical behaviour therefore making definitive diagnosis and treatment difficult \(^8\). These are basically considered as benign neoplasm with high malignant potential. The chances of metastases is approximately one half of the patients with malignant hemangiopericytoma. the site most frequently affected site being the lungs followed by bones, liver, and lymph nodes. The spread of tumor is predominantly hematogenous.

Based on radiological examination, they may either be interpreted as lytic or focal sclerosis or may also show honeycomb or reticular pattern. Sometimes, cortical erosion can be seen which may signify the presence of malignancy\(^5\).

The standard treatment for HPC is surgical excision. Optimal treatment is wide excision with a margin of 1cm of normal tissue. More extensive surgery is required for tumours with malignant characteristics. The role of radiotherapy in the treatment of HPC is controversial because these tumors are considered radioresistant. However, radiotherapy can be used as an adjunctive treatment modality for recurrent tumors, postoperative surgical fields, or treatment of inoperable metastases. According to Staples et al \(^10\) post-operative radiation therapy contributes to decreasing the recurrence ratio and reported that hemangiopericytomas are relatively radiosensitive. H. Hiraumi et al. \(^11\) in their study, used radiation therapy as their initial treatment because of the poorly controlled diabetes mellitus and rapid growth of the tumour. The tumor showed remarkable radiosensitivity and the patient did not accept wide surgical excision. They concluded that the Hemangiopericytomas have a variable clinical nature, and some of them may be highly radiosensitive and radiation therapy can be a treatment of choice in the management of aggressive tumors.

In our case, a case-report of hemangiopericytoma of the palate is presented. Management of the tumor was done by wide surgical excision. Radiotherapy was not advised because there was no lymphadenopathy. The patient was under observation and regular follow-up for 24 months after surgery and showed no recurrence.

We consider that, in tumors without metastasis, wide surgical excision and not enucleation, is generally enough for management of hemangiopericytomas of the head and neck region and the choice of radiotherapy should be reserved for recurrent, unresectable and metastatic cases.
Conclusion:
Hemangiopericytoma, though a rare, controversial tumor, should be considered in the differential diagnosis of apparently benign lesions of the oral cavity. There is a need for carefully planned treatment and long-term follow-up of the patient because of the potential malignant behaviour of the tumor. Wide local excision being the gold standard treatment for this tumor, studies are still needed to further deeply explore the significance and accuracy of adjuvant therapy in the setting of a high recurrence rate of such a vascular tumor.

References:
1. Michael s. Goldwasser, and joseph l. Daw. hemangiopericytoma of the palate: case report J oral maxillofac surg. 1990; 48:211-215.
2. Ceylan A et al. Hemangiopericytoma of the hard palate. Dentomaxfac Radiol 2008;37(1), 58–6.
3. Stout AP, Murray MR. Hemangiopericytoma: A vascular tumor featuring zimmermann’s pericytes. Ann Surg 1942;116(1):26-33.
4. Shobha BV, Shivakumar BN, Reddy S, Dutta N. Sinonasal hemangiopericytoma: A rare case report with review of literature. J Oral Maxillofac Pathol 2015; 19:107.
5. Kendre P, Kataria P, Patel AA, Gaurav L, Dalsaniya S. Hemangiopericytoma of supraglottis: A rare case report and review of literature. J Can Res Ther 2019; 15:729-32.
6. Raghani N, Raghani MJ, Rao S, Rao S. Hemangiopericytoma/Solitary fibrous tumor of the buccal mucosa. Ann Maxillofac Surg 2018; 8:151-3.
7. S.M. Florence, C. Willard, C.W. Palian, Hemangiopericytoma of the Buccal Region: A Case Report. J Oral Maxillofac Surg 2001; 59:449-453.
8. P. S. Kothari, M. Murphy, G. L. Howells, D. M. Williams. Hemangiopericytoma: a report of two cases arising on the lip. Br J Oral Maxillofac Surg 1996;34,454-456
9. Enzinger FM, Smith BH. Hemangiopericytoma. An analysis of 106 cases. Hum Pathol 1976; 7:61-82.
10. Staples JJ, Robinson RA, Wen BC, Hussey DH. Hemangiopericytoma – the role of radiotherapy. Int J Radiat Oncol Biol Phys 1990; 19:445–51.
11. H. Hiraumi et al. Radiosensitive hemangiopericytoma in the soft palate. Auris Nasus Larynx 2002; 29:95–97.