CASE REPORT

Sinonasal hemangiopericytoma: A rare case report with review of literature

BV Shobha, BN Shivakumar, Santhosh Reddy, Neerav Dutta
Department of Oral and Maxillofacial Pathology, Maitri College of Dentistry and Research Centre, Anjora, Durg, Chhattisgarh, India

Address for correspondence:
Dr. Shobha BV,
Maitri College of Dentistry and Research Centre,
Anjora, Durg - 491 001, Chhattisgarh, India.
E-mail: shobhabvs@gmail.com

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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 22-year-old female, who presented to our department with a tender swelling in maxillary anterior region and the mass was well-circumscribed, sessile and soft on palpation. The skin over the tumor was intact and normal. The tumor was completely removed with wide surgical resection. The histopathological staining supported the diagnosis of HPC, this was further confirmed by immunohistochemistry (IHC) in which CD99 showed strong positivity.

Key words: Anterior maxilla, CD99, hemangiopericytoma, staghorn pattern

INTRODUCTION

Hemangiopericytoma (HPC) is a soft tissue tumor arising from Zimmermann's pericytes, which are modified smooth-muscle cells in the periphery of blood vessels. These pericytes are located outside the reticulin sheath of the endothelium.[1,2]

Pericytes which are small, have shape such as oval or spindle-shaped cells lining the capillaries.[3]

HPC can occur in any age group and there is no sex predilection. The HPCs exhibit an unpredictable biologic behavior. Malignant forms show necrosis, cellular pleomorphism, high proliferation index and mitoses >4 per 10 high power fields. The absence of necrosis, cellular pleomorphism and mitoses <4 per 10 high power fields does not necessarily indicate benign nature; infact, tumors with benign histological appearance have been reported to metastasize.[1]

HPC is an uncommon mesenchymal tumor, accounting for 1% of all blood vessel-related neoplasms and approximately 3% of all soft tissue sarcomas. HPC has a predilection for the long bones, pelvis and scapula; but, 15% occur in head and neck region, incidence of sinonasal HPC is less than 1%. Origin in oral cavity is less common. In an analysis by Brockbank, from 1949 to 1979, only 35 cases were reported in the oral cavity.[1,4,5]

HPC is a rare tumor of adult life mostly occurring in 5th decade and is very uncommon in children and accounts for only 10% of cases.[6]

The architectural pattern of HPC can be seen in other mesenchymal neoplasms. The diagnosis of HPC is one of the exclusions and relies on the presence of characteristic histological features. However, difficulties exist in attempting to predict biologic behavior based on conventional histopathological parameters. As sarcomas, HPCs are graded based on histologic and biologic parameters. However, the pericytes, in which they originate, possess characteristics of both smooth muscle and endothelial cells. Differentiating these from other cell types is often challenging. Accordingly, the diagnosis of HPC is made on the basis of distinct architectural patterns exhibited histologically.[7,8]

The diagnosis of HPC is based on the following criteria: On immunohistochemical analysis, the tumor cells are negative for desmin, S100 protein, α-smooth muscle actin and cytokeratin (CK); and are intensely positive for vimentin and are focally positive for CD34.[1,9]

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Prior to the routine use of immunohistochemistry (IHC) in the diagnosis of HPC, misinterpretation for synovial sarcoma was common. However, unlike HPC, synovial sarcomas are often immunoreactive for both keratins and epithelial membrane antigen (EMA). Electron microscopy is also helpful in separating the two tumors, as epithelial differentiation is identified in some synovial sarcomas studied ultrastructurally.[10]

Increased mitotic activity, a higher cell density, an appearance of undifferentiated cells; and presence of necrotic and hemorrhagic areas in the tumor tissue are the major features seen during malignant transformation. HPC shows cytogenic abnormalities.[11]

Depending upon the size and location of the tumor, the HPC generally present as painless mass, but may have symptoms.[12]

We report here a case of HPC of the anterior maxilla in a 22-year-old female along with the literature review of HPC.

CASE REPORT

A 22-year-old female reported with the complaint of swelling at the left side of anterior maxilla, since 3 months [Figure 1]. Intraoral examination revealed left buccal and palatal swelling extending upto the posterior part of the hard palate. The swelling was approximately 2 × 3 × 1.5 cm in size. Overlying mucosa appeared normal in color. On palpation, the lesion was sessile, the growth pattern was intermittent and the surface appeared smooth. The lesion on palpation appeared soft and was fixed to the underlying structures. No engorged vessel or discharging sinus was noted. Regional lymphadenopathy was not seen; also, there was no history of pus discharge or foul smell from the site. Oral hygiene status of patient was satisfactory and all the teeth were periodontally sound [Figure 2].

Panorex X-ray showed a poorly circumscribed lesion with radiopaque soft tissue mass which displaces neighboring teeth 22 and 24 palatally with 23 missing with no root resorption seen in both the displaced teeth [Figure 3].

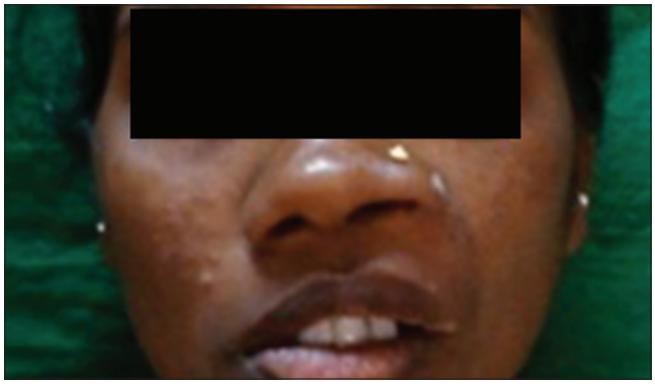

Figure 1: Swelling seen in the left anterior maxillary region

Incisional biopsy was done and diagnosis of sinonasal HPC was made.

On macroscopic examination, the lesion was roughly oval (0.8 × 0.5 × 0.4 cm). Sectioning revealed a variegated cut surface with yellow and grey areas. Firm consistency was seen.

In the present case, following histopathological features were evident. The tumor was composed of tightly packed cells surrounding thin-walled blood vessels. The tumor cells were round to ovoid in shape with well-defined cell boundaries. Areas of hyalinization with numerous collapsed thin-walled blood vessels within them were evident. Typically these blood vessels, lined by flat endothelial cells are arranged in a "staghorn pattern". Cytological atypia is minimal, presence of necrosis is evident, margins are well-defined and infiltrative pattern is not seen. Mitotic activity is sparse. Clearly giving a definitive picture of low-grade HPC [Figures 4-6]. Additional confirmation with IHC was done and the results showed that CD99 staining was positive and CD34 was scattered cell positive in which the vasculature was highlighted. Whereas, CK, desmin and S100 were negative.

The differential diagnosis in this case includes solitary fibrous tumor and poorly-differentiated synovial sarcoma. Normal endothelium lines the blood vessels in contrast to malignant angiosarcoma, where the malignant tumor lines the vascular spaces. Histologically various other tumors may show a vascular pattern which resembles HPC. Mesenchymal chondrosarcoma, fibrous histocytoma and synovial sarcoma should be considered as differential diagnosis. The negative staining with CK rules out synovial sarcoma. Solitary fibrous tumors reveal strong CD34 positivity (>90%) and are the defining features of these tumors. In most situations, the diagnosis of HPC is one of exclusion; however, in the present case, HPC was diagnosed based on clinicopathological correlation [Figures 7 and 8].

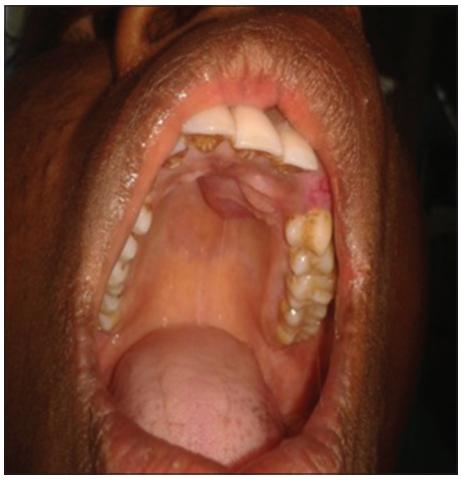

Figure 2: Intraoral view showing left buccal and palatal swelling

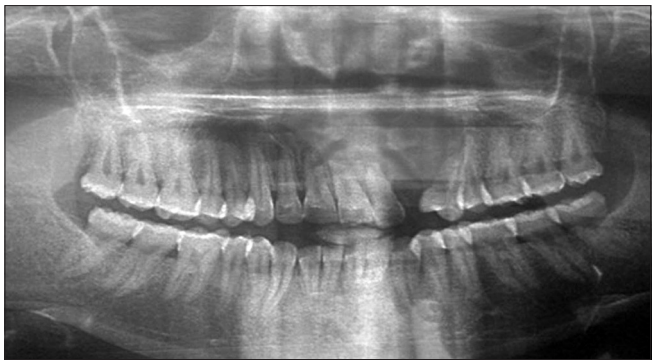

Figure 3: Orthopantomogram showing a poorly circumscribed lesion showing radiolucency with 23 missing

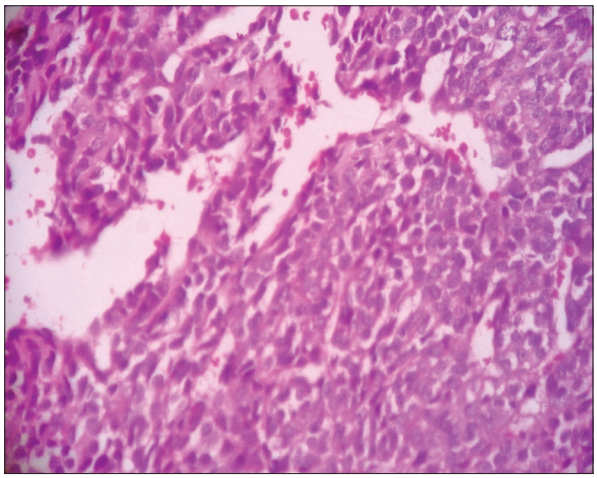

Figure 5: Photomicrograph showing typical staghorn pattern of vascular channels, the blood vessels are lined by flat endothelial cells(H& E stain, x400)

The treatment was done with complete wide surgical maxillectomy. The patient was discharged after full recovery. Patient is on regular follow up since last 1 year. No sign of recurrence or metastasis have been noted so far.

DISCUSSION

The tumor was first described by Stout and Murray in 1942 (Stout and Murray, 1942), HPC is a soft tissue tumor derived from mesenchymal cells with pericytic differentiation (Enzinger and Weiss, 1995). Disease can be both benign and malignant. Two types of HPC have been described; namely, infantile HPC and adult HPC disease. Although the infantile variety is mostly described with the adult type; this type deserves a special mention because of its different histological presentation and clinical behavior.[6]

HPC is a one of the rare mesenchymal tumor that occurs as a localized tumor mass with diameter ranging from 1 to 20 cm. Based on the medical history and clinical findings, patient's various etiological factors have been suggested as hypertension, hormonal or metabolic imbalance and trauma;

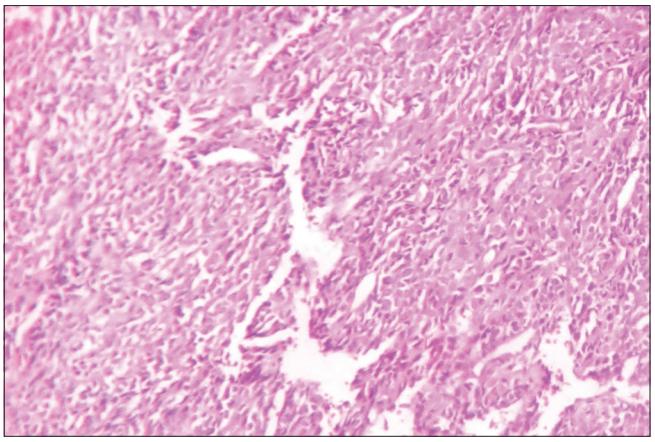

Figure 4: Photomicrograph showing staghorn pattern and blood vessels are lined by flat endothelial cells (H&E stain, x100). H&E: hematoxylin and eosin

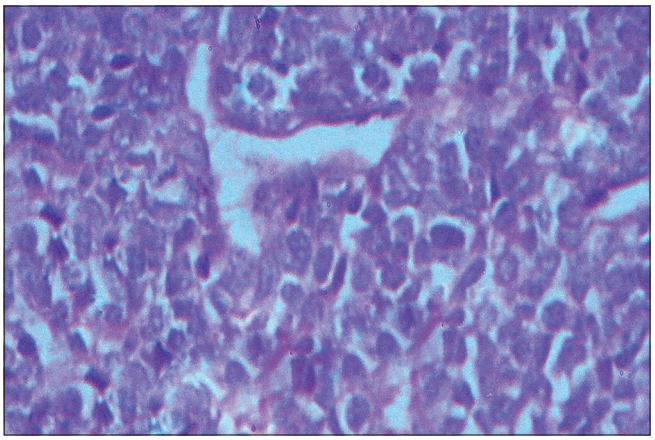

Figure 6: Photomicrograph shows network of capillary vessels with a staghorn pattern, where lumen is lined by flat endothelial cells. The tumor cells present a pale cytoplasm with round or ovoid nuclei(H&E stain, x400)

but the etiology of HPC is unknown. In HPC, cytogenetic abnormalities are seen in most of the cases. Most HPC are mostly near diploid and breakpoints in 12q13, 12q24 and 19q13 seem to be very common with recurrent t (12;19) (q13;q13) translocation (Mandahl *et al*., 1993; Mitelman *et al*., 2002; Hallen *et al*., 2002).[1,6,11]

HPC constitutes only 3–5% of all soft tissue sarcomas and about 1% of all vascular tumors. The head and neck incidence is 15–30% and it is mostly seen in adults. In the head and neck region, it is mostly found in the parapharyngeal space, masticator space, orbit, nasal cavity, oral cavity, jaw, parotid gland and jugular foramen. Brockbank analyzed a series of 35 cases in the oral cavity which included: Tongue (nine cases), upper jaw bone (five cases), lips (four cases), buccal region (three cases), gingiva (three cases), parotid gland (one case) and multifocal lesions (one case).[1,13,14]

HPC have been described in all age groups; with more than 40% occurring in the 5th and 6th decades of life, but as in

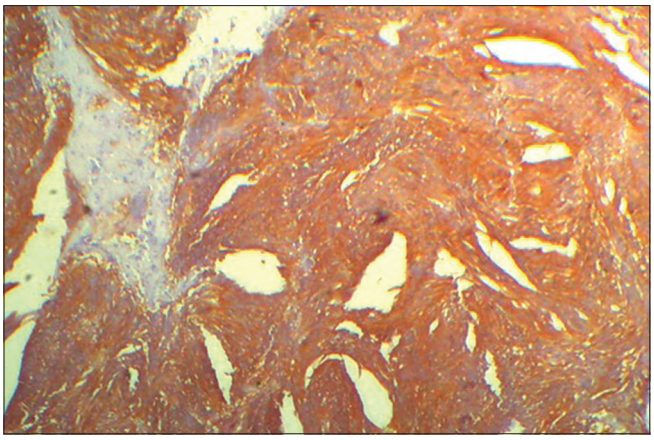

Figure 7: Tumor showing positive reactivity with CD34 typically around vasculature (IHC stain, x40)

present case it can occur in 2nd and 3rd decade of life as well. Only 10% of HPC occur in children. The tumor has no sex predilection. Clinically the lesion appears firm, apparently circumscribed, often nodular and may or may not exhibit redness which is an indicative of their vascular nature. As in present case, redness was not seen and the swelling was apparently soft rather than firm. Painless enlarging mass is the general mode of presentation as was seen in present case. Majority of tumors grow rapidly and are therefore of short duration as in our case in which the growth was intermittent with rapid growth and recurrence was seen.[5,6]

Radiographic features of HPC vary drastically and resemble a malignant bone lesion. Radiographically, the lesion may be well circumscribed and sometimes appears to be radiopaque with displacement of surrounding structures. As in present case, a well-defined radiopacity was evident with the simultaneous displacement of nearby teeth palatally. Our finding was in accordance with the previous literature.[5,13]

Benign and malignant variant of this tumor have been reported histologically. In 1975, Mc Master mentioned the histopathological criteria for differentiating the benign and malignant HPC and later it was modified by Enzinger and Smith in 1976. The biological behavior of this tumor is quite unpredictable, so histologically benign tumors can sometime behave aggressively and will have the capacity to metastasize.[15]

Histological findings of these tumors are classified as low-, intermediate, or high-grade, which are based on cellular pleomorphism, mitosis and cellularity. Tightly packed cellular areas surrounding thin-walled branching blood vessels are present in tumors. The tumor cells are of variable small, ovoid to spindle shaped showing ill-defined cell boundaries. Blood vessels are collapsed, lined by flat endothelial cells which are arranged in a characteristic "staghorn" pattern. Normal endothelium lines the blood vessels in contrast to malignant angiosarcoma, where the malignant tumor lines

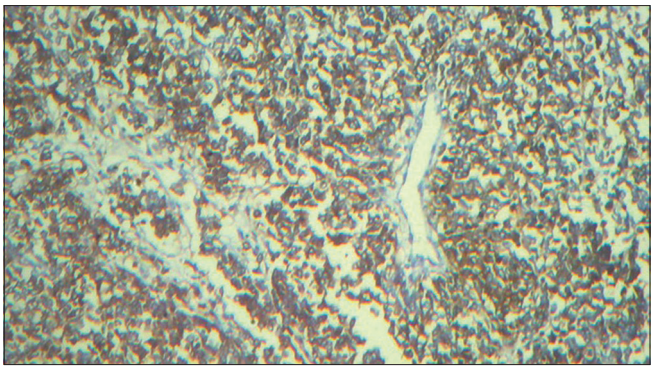

Figure 8: Tumor showing positive reactivity with CD 99 staining (IHC stain, x100)

the vascular spaces. Histologically, various other tumors may show a vascular pattern which resembles HPC. Mesenchymal chondrosarcoma, fibrous histocytoma and synovial sarcoma should be considered as differential diagnosis.[1,5,6]

In the present case, similar histopathological features were evident. The tumor is composed of tightly packed cells surrounding thin-walled blood vessels. The tumor cells are round to ovoid in shape with well-defined cell boundaries. Areas of hyalinization with numerous collapsed thin-walled blood vessels within them are evident. Flat, endothelial cells lines the blood vessels which are arranged in a "staghorn pattern". Cytological atypia is minimal, presence of necrosis is not evident, margins are well-defined and infiltrative pattern is not seen. Mitotic activity is sparse; giving a picture of low-grade HPC.

The differential diagnosis between HPC and other tumors exhibiting vascular proliferation requires IHC. Vimentin is the only marker that is consistently expressed in HPC and CD34 is also found to be positive in the tumor cells. Vascular markers stain only the endothelial cells of the blood vessels. The tumor that most closely resembles HPC is the solitary fibrous tumor, whose cells also stain for vimentin and CD34. In the present case, CD34 was found positive, particularly around the blood vessels. Contrary to this, solitary fibrous tumors reveal strong CD34 positivity (>90%) and is the defining feature of these tumor. Other markers such as CK, desmin and S100 were negative. The negative staining with CK rules out synovial sarcoma.[1,16]

The treatment of choice for HPC is wide surgical excision, with preceding ligation of the vascular bundle that nourishes the neoplastic tissue, thus achieving reduction of the size of the neoplasm as well as its removal. Chemotherapy and immunotherapy have been considered for control of malignant HPC and metastatic disease. Radiation therapy has usually been reserved for unresectable and recurrent tumor. The efficacy of radiation therapy is doubtful because HPC is considered to be radioresistant, but radiation therapy can be useful in the treatment of aggressive HPC and incomplete resections.[1,5,6,11]

Prognosis of HPC is usually favorable and depends on mitotic activity in tumor. Espat *et al*., reported 93% and 86% mitotic activity with 2- and 5-year overall survival rates, respectively. Both of them reported that incidence of metastases varies from approximately 10 to 60%, depending on the diagnostic criteria and the therapy. According to findings by Enzinger and Smith, more than two-third of the cases which eventually metastasize mostly develop local recurrences before metastasis. HPC cases with high-grade malignancy have been reported with lymphnode involvement; bone, pulmonary and hepatic metastases.[1,5,11,17]

CONCLUSION

HPC is an uncommon vascular tumor in which the biologic behavior is difficult to predict when based solely on conventional histological parameters. Features suggestive of a lesion with an increased risk for subsequent recurrence or metastasis include increased cellularity, necrosis, hemorrhage and increased mitotic activity. These features can be supplemented by determining the proliferation index using immunohistochemical techniques. Recommended treatment is wide surgical resection. Long-term follow-up is mostly necessary in patients even after radical resection because recurrence or metastases may be delayed by many years. Radiotherapy and chemotherapy can cause tumor regression and are not suggested as primary treatment.

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