

Original Research Paper

Hemangiopericytoma of the Lower Lip Distinct and Rare Entity

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ABSTRACT

Hemangiopericytoma is considered as a rare entity, arising from mesenchymal cells with pericytic differentiation. Hemangiopericytoma is most commonly seen in adults the tumor is extremely rare in the head and neck region. Surgical resection remains the mainstay treatment. Late relapses may occur and require long-term follow-up. We have a case of 63 years old male patient present with lower lip mass, which was excised and the histopathology diagnosis came a hemangiopericytoma.

Key words: Hemangiopericytoma , rare, lip, favorable prognosis.

INTRODUCTION

Hemangiopericytoma is an uncommon tumor of cells of mesenchymal origin that partially surround the endothelial cells of capillaries and small veins. The mean age of presentation is usually 63 years, but has a wide age range from 5-86 years, it has a slight female predominance , and may be found at any location. 40% of tumors are found in the subcutaneous tissue, others are found in the deep soft tissues of the extremities or extra- compartmentally in the head and neck region, (especially the orbit and oral cavity), thoracic wall, mediastinum, pericardium, retroperitoneum. Other described locations include the meninges, spinal cord, periosteum as well as organs such as the salivary glands, lungs, liver, gastrointestinal tract, urinary bladder, etc. Recurs locally (18%), no metastases and death due to disease are rare. We report a case of 63 years old male patient present with lower lip mass , which was excised and the histopathology diagnosis came a hemangiopericytoma.

CASE REPORT

A 60-year-old male was referred to and examined at a private clinic after the of a gradual swelling on his lower lip which had started to grow gradually in the past one year and it grew slowly, which gave the patient an uncomfortable appearance and difficulty in eating, while at the same time experiencing difficulty speaking. The aforementioned patient's medical history was unremarkable, and because of this, he sought medical advice in the ear-nose-throat (ENT) clinic. On examination, there was a soft tan, brown movable mass in the lower lip about 3 cm in maximum diameter, a biopsy of the tumor was performed, and diagnosed in the pathology laboratory as hemangiopericytoma. The biopsy specimen had a high cell density and infrequent mitotic figures/HPF. Resection was scheduled and the patient was admitted to

the Hospital and the tumor was resected under general anesthesia. A 5-mm safe margin, the defect was covered with artificial dermis graft.

The patient was discharged. Since that time he has remained under observation. Till date, there has been no evidence of recurrence or metastasis. Macroscopically, shows a tumor with 5-mm surgical margin was well-circumscribed (3,4,5,12) masses with a yellowish tan cut surface and a fleshy consistency with a variable area of Haemorrhage but no necrosis tumor size is 4 cm in maximum diameter. Microscopically, shows the solid cellular areas consistent presence of numerous, variably ectatic and thin-walled branching vessels having a staghorn configuration. Tumor cells are closely packed, spindle-shaped to round, of uniform size, with small amounts of pale or eosinophilic cytoplasm with indistinct margins and small, bland hyperchromatic nuclei. rare mitosis are seen about 2/10 HPF

. No necrosis .Figure 1

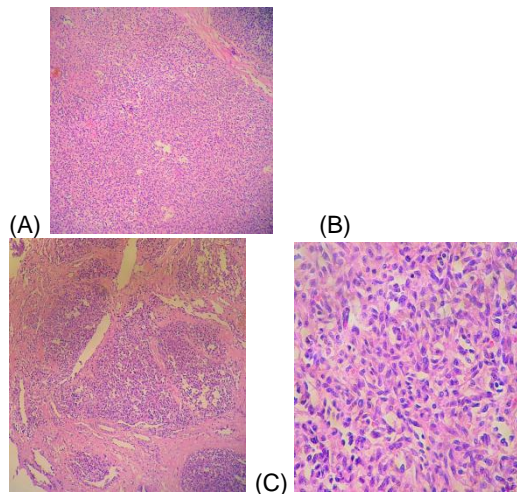


Figure 1 Histological findings. (a) Lower magnification shows the dense, cellular rich tumor cells arranged around the numerous thin-walled vessels (HE stain, original magnification X100). (B) Within tumor nests, thin-walled branching vessels, stag-horn configuration are observed (HE stain, original magnification X200)(C) higher power examination show spindle to round closely backed cell with pale cytoplasm and bland hyperchromatic nuclei (HE stain original power x400)

Immunohistochemistry

shows fair positivity for CD34 and SMA , Figure 2.

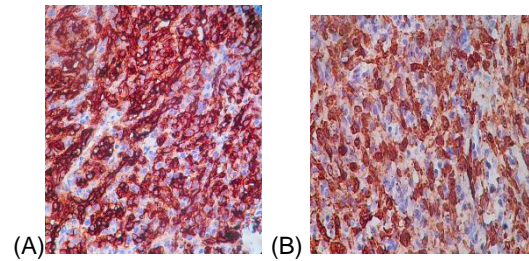


Figure 2

(A) Immunohistochemical staining shows tumor cells with strong positivity for CD34 (original magnification x400). (B) Immunohistochemical staining shows tumor cells with diffused positivity for SMA (original magnification X400).

DISCUSSION

Hemangiopericytoma is a tumor that arises from pericytes,(1,3,4,5), surrounding capillary vessels. It can arise anywhere capillary vessels are found, commonly seen in deep soft tissue, particularly pelvic retroperitoneum. It can be seen in the head and neck particularly in the oral cavity , tongue, lip, and gum , a distinct variant is seen also in the Sinonasal area A smaller proportion of cases arise in the proximal limbs or limb girdles. Histologically comparable lesions also occur in the meninges. In the 2002 World Health Organization(WHO) classification 12 , it is not categorized as neither benign nor malignant but is regarded as a tumor with potential low malignancy. In general, hemangiopericytoma is believed to occur at an equal rate in both sexes (11,14,16) , and all age groups.5–80 with a mean age group of 60 years as in our patient.

The clinical symptoms usually present itself as a slowly growing mass which, in the abdomen, may cause intestinal or urinary symptoms. Occasional cases, similar to SFT, are associated with hypoglycemia due to secretion of insulin-like growth factor . Macroscopy HPC tends to be well-circumscribed (15,18,20), masses with a yellowish or tan cut surface and a fleshy or spongy consistency. Large vessels may be evident on the cut surface. Haemorrhage is common, but necrosis is infrequent. Tumor size is variable, but most cases are 5-15 cm in maximum diameter.

Histopathology, the HPC closely resembles the cellular areas of SFT, albeit, with the presence of numerous, variably ectatic or compressed, thin-walled

branching vessels often having a staghorn configuration. Tumor cells are usually closely packed, spindle-shaped to round, of uniform size, with small amounts of pale eosinophilic cytoplasm with indistinct margins and small, bland often vesicular nuclei(11,15,13). Cytological pleomorphism is generally not a feature.

The mitotic rate is highly variable. Some cases contain a prominent adipocytic component (such cases are known as lipomatous HPC). These lesions also often show varying cellularity and are increasingly regarded as a variant of SFT. Tumors which very often were classified as HPC in the past include solitary fibrous tumour, monophasic synovial, sarcoma infantile myofibromatosis, myopericytoma, infantile fibrosarcoma, deep fibrous histiocytoma and mesenchymal chondrosarcoma.

Immunohistochemistry.

The HPC shows fairly positivity for CD34 and CD99, both of which are widely expressed in fibroblastic tumors. Endothelial markers are negative. Ultrastructure most of the lesions reported as HPC have shown only undifferentiated spindle cell or fibroblastic features. Convincing evidence of true pericytic differentiation(6,7,8,14), is not seen. Genetics study of the few cytogenetically investigated HPCs, located in the lung, tongue, brain, cerebellum, soft tissues, and intraabdominal, have been reported. The vast majority of cases have had near or pseudodiploid karyotypes with the number of aberrations ranging from one to more than 20. The chromosome aberrations are quite disparate, but breakpoints in 12q13-15 and 19q13 have been identified in almost half of the cases and one-fourth of the cases, respectively.

In two cases, there was a balanced(12;19)(q13;q13), in one case as the sole anomaly. Among the genomic imbalances, losses are predominating. Recurrent imbalances include loss of segments in 3p, 12q, 13q, 17p, 17q, 19q, and the entire chromosome 10, and gain of 5q sequences. At least 70% or more of HPCs pursue a benign clinical course, (15,17,18) while the remainder is malignant. Histological criteria for malignancy are cut of 4 or more mitotic figures per 10 HPF is the single feature most worrisome for malignancy. The presence of necrosis or nuclear pleomorphism, particularly in the context of a tumor >5 cm in diameter may also portend malignant behavior.

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