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Review Article
Hemangiopericytoma: A Case Report and Mini-Review

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Keywords

Hemangiopericytoma, vascular neoplasm, pericytes.

Abstract

The term hemangiopericytoma was first used by Stout and Murray in 1942 describing a tumor which is distinguished histologically from other types of vascular neoplasm by its proliferation of pericytes. Hemangiopericytoma is a rare neoplasm that was originally described as a vascular tumor derived from the pericytes. They account for 2-3% of all soft tissue sarcomas in humans and they occur mainly in the musculoskeletal system. 15-30% of all hemangiopericytomas occur in the head and neck region. Only 5% are located in the sinonasal region, where they display a more benign behavior than in other parts of the body. Herein, we are presenting an extremely rare case report of hemangiopericytoma in a 32 year old male patient with a brief overview regarding its epidemiology, macro- and microscopical characteristics, the clinic-pathological findings and the treatment of this extremely rare vascular neoplasm.

Introduction

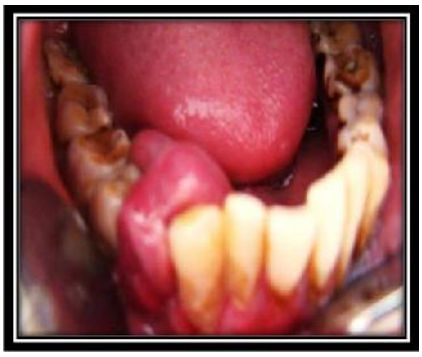
Hemangiopericytomas are distinct neoplasms of vascular origin which were first affirmed by Stout and Murray in the year 1942. These tumors most often show benign biologic behavior, however, very rarely, they may also present with malignant characteristics. These tumors are very rare in their occurrence with only 2-3 % of all soft tissue neoplasms being comprised of hemangiopericytomas. Moreover, of all the reported hemangiopericytomas, about 15-30 % occur in the head and neck region. 5% of these tumors are located in the sinonasal region where they possess less aggressive behavior in comparison with tumors occurring elsewhere in the body. [1]
The word hemangiopericytoma originates from the ancient Greek words: haema (from Ancient Greek αἷμα, haima, meaning "blood"), angio (meaning blood vessel), oma (meaning tumor or neoplasm), peri (meaning "about" or "around", "enclosing" or "surrounding", and "near") and cytoma (referring to the cells surrounding the blood vessel walls). Thus, hemangiopericytomas can develop wherever pericytes are present. The tumor holds specific significance due to its potential for malignant behavior. Some of the known sites for tumor development are the pelvic region, retroperitoneum and along the extremities, however, primary bone localization is not very common. Tumor manifestation signs usually depend on tumor's location, size and potential for malignancy.[2] The following case report describes a very rare case of hemangiopericytoma located in the oral cavity & also takes a glance on relevant literature about the same.

The oral hemangiopericytomas are fast growing tumors, with a characteristic bluish red color. There is no age or gender predilection for these tumors. However, the lesions are rare reported before 20 years and after 70 years of age. Their consistency varies from being soft to rubbery and most commonly, the lesions are not associated with pain.

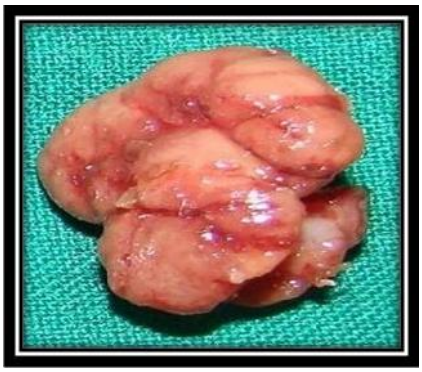
The lesions also are often well-delineated from the surrounding mucosa. Chromosomal translocations t (12;19) and t (13;22) have been observed in lesional cells.[2] Tumors seen in perivascular areas especially deep seated tumors in muscles are larger tumors. The smaller tumors are usually benign. In contrast, the larger lesions are more aggressive and are feared for their malignant behavior, thereby, requiring more aggressive treatment with wide resection followed by postoperative radio and chemo-therapies.[3]

Case Report

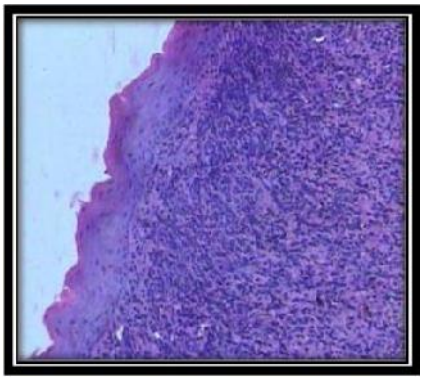
A 32-year old male patient visited the Department of Oral Medicine and Radiology/Outpatient Department of our College with a chief complaint of swelling in relation to the lower right back tooth region in the lingual aspect since 8 months. The history revealed that the swelling had started insidiously, not preceded by trauma, which was initially smaller in size and steadily increased in size since its onset. Patient had experienced mild discomfort causing difficulty during speech and mastication and it was not associated with any history of pus discharge. Patient was operated for the same swelling along with extraction of grossly carious 44 in a private set-up 3 months back. Lesion recurred and patient visited the same clinician again for the similar complaint. Medical, family, and personal histories were not relevant. General physical examination revealed no abnormalities. Extraoral examination revealed no gross asymmetry. No regional lymphadenopathy was evident. Intraoral examination revealed a solitary, painless, sessile, oval swelling in the mandibular lingual sulcus, measuring about 2x3 cm in its greatest dimensions, extending anteroposteriorly from the mesial aspect of 43 till the mesial aspect of 45 and superoinferiorly from the occlusal surface of teeth in the affected region till the attached gingivae in the same quadrant. The overlying mucosa was red in color. It was firm in consistency with well defined borders, lobulated in shape and smooth in texture (Fig.1). On palpation, it was fixed to the underlying gingiva and it slightly bled on probing. Clinical differential diagnosis included fibroma, peripheral giant cell granuloma and pyogenic granuloma. Radiological investigations revealed no distinct radiological characteristics. Routine hematological investigations revealed normal values. Under aseptic conditions, the lesion was surgically excised (Fig.2). The H&E stained sections showed parakeratinized stratified squamous epithelium with corrugated surface layer and underlying connective tissue stroma (Fig.3). The underlying connective tissue stroma showed lesional tissue which was highly cellular in nature with numerous vascular spaces lined by endothelial cells (Fig.4). Irregular branching of vascular spaces revealed characteristic ‘stag horn’ pattern (Fig.5). Surrounding these vascular spaces, there were proliferations of tightly packed oval and spindle cells showing indistinct cytoplasmic borders with moderate amount of cytoplasm and hyperchromatic nuclei (Fig.6). The histopathological impression was suggestive of hemangiopericytoma.



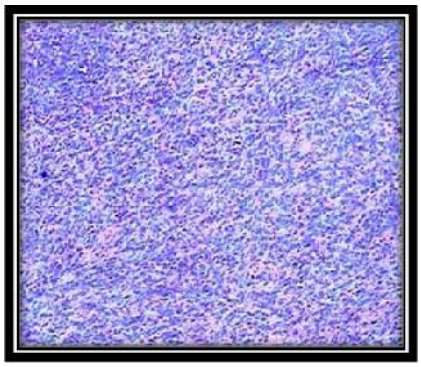
(Fig.1) revealing a solitary, sessile, oval shaped growth in the mandibular lingual sulcus, measuring about 2x3 cm in its greatest dimensions, extending anteroposteriorly from the mesial aspect of 43 till the mesial aspect of 45 and superoinferiorly from the occlusal surface of teeth in the affected region till the attached gingivae in the same quadrant



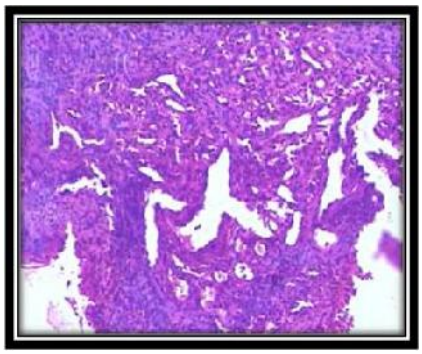
(Fig.2) revealing the excised specimen



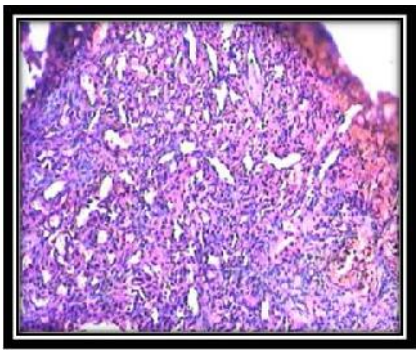
(Fig.3) revealing the H&E stained section showing parakeratinized stratified squamous epithelium with corrugated surface layer and underlying connective tissue stroma



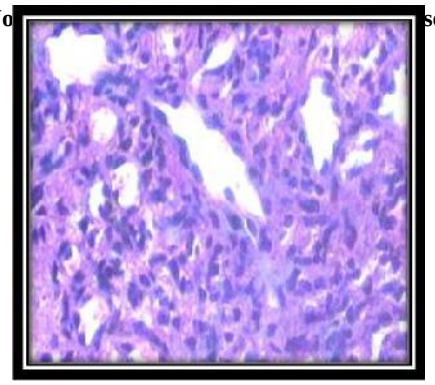
(Fig.4) revealing the underlying connective tissue stroma showing lesional tissue which was highly cellular in nature with numerous vascular spaces lined by endothelial cells



(Fig.5) revealing irregular branching of vascular spaces in the characteristic ‘stag horn’ pattern



(Fig.6) revealing proliferations of tightly packed oval and spindle cells showing indistinct cytoplasmic borders with moderate amount of cytoplasm and hyperchromatic nuclei around the vascular spaces



(Fig.7) revealing reticulin staining demonstrating lesional vessels lined by a single layer of endothelial cells with the pericytes covering the basement membrane of blood vessel

Discussion

Hemangiopericytoma is a rare neoplasm that was originallyescribed as a vascular tumor derived from the pericytes. Clinically, the lesion may be either sessile or pedunculated, and may demonstrate a surface lobularity with a telangiectasic appearance. Intraosseous cases have also been reported.[2,3] Hemangiopericytomas have been reported to occur in all age groups however, majority (40%) of the cases occur in the 5th and 6th decades.[3,4]

Hemangiopericytomas do not have any specific radiological characteristics. They may be either lytic or may represent focal sclerosis. They may also show a honeycomb or reticular pattern. Tumors may also cause cortical erosion, which alarms the presence of a malignancy. Along with routine radiography, even CT scans and MRIs are not contributory in diagnosis. However, they are beneficial to differentiate benign tumors from those which are malignant. They are although also significant to determine the extent of the tumor. Angiography of the tumor reveals spider-like, radially branching vessels.[3]

Histology of these tumors is although very characteristic. It depicts numerous vascular channels with plump endothelial cells. The stroma surrounding the vascular channels consists of proliferation of tightly packed ovoid and spindle shaped cells with hyperchromatic nuclei and a moderate amount of cytoplasm. The cells do not have distinct cytoplasmic borders. The tumor cells are seen surrounding the proliferating vascular channels. The irregular vascular channels show branches of varying sizes that give the characteristic 'staghorn' appearance to the histology of these tumors. Older lesions are less aggressive and they have less cellularity in comparison to the younger lesions. They may possess a large mucoid interstitial appearance which can appear as the one in mucoid type of lipoma or that of liposarcoma of myxoid type. Rarely, these tumors may also show focal areas of cartilage production and in such cases, have to be cautiously differentiated from mesenchymal chondrosarcomas.[1,4]

On gross examination, hemangiopericytoma appears grayish-white in appearance & may be well circumscribed. The appearance is much less hemorrhagic compared to the endothelial tumors. The consistency varies from being solid to spongy, friable or granular. One variant of this tumor is congenital hemangiopericytoma which is also known as infantile hemangiopericytoma. Oro-pharyngeal mucosa is the predominant location for this very rare lesion. This tumor variant most often is present at birth, occurs more than one in number, and at various sites along the mucosa. It shows faster growth rate after birth. Recurrence rate is high for this variant of the tumor, although they are less prone to develop metastasis as is reflected in the literature.[4]

The histogenesis of hemangiopericytomas revolves around pericytes. Pericytes are a normal component of blood capillaries. These are contractile cells that are zipped around the endothelium of small blood vessels all over the vasculature. Thus, hemangiopericytomas can occur anywhere in the body. Rouget cells or mural cells are other names for pericytes. They are found embedded in the basement membrane & provide communication with the endothelial cells of blood vessels. There is dual mode of communication both via direct physical contact and via paracrine signaling. Pericytes help in regulation of blood flow along the capillary walls and are also involved in washing out of cellular waste by the process of phagocytosis. They stabilize and monitor the maturation of endothelial cells. Apart from their role along the capillary bed, pericytes are also a significant component of the neurovascular unit, which includes endothelial cells, astrocytes and neurons.[2] Pericytes and endothelial cells lie in close proximity to each other and thereby it is difficult to identify both of these cell types. However, these two cell types can easily be differentiated from one another on the basis of cytomorphology of the cell species with pericytes having prominent round nuclei compared to the flat elongated nuclei of the endothelial cells.[3] Ultrastructurally, pericytes have numerous finger- like projections on their surface that are wounded around the capillary walls and help in directing the blood flow. [2,4]

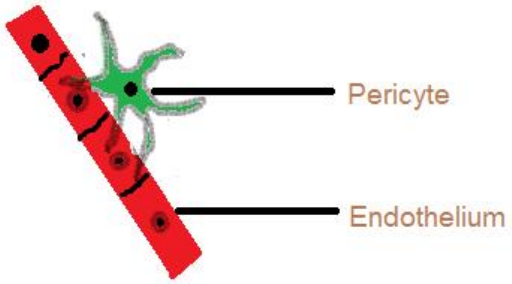
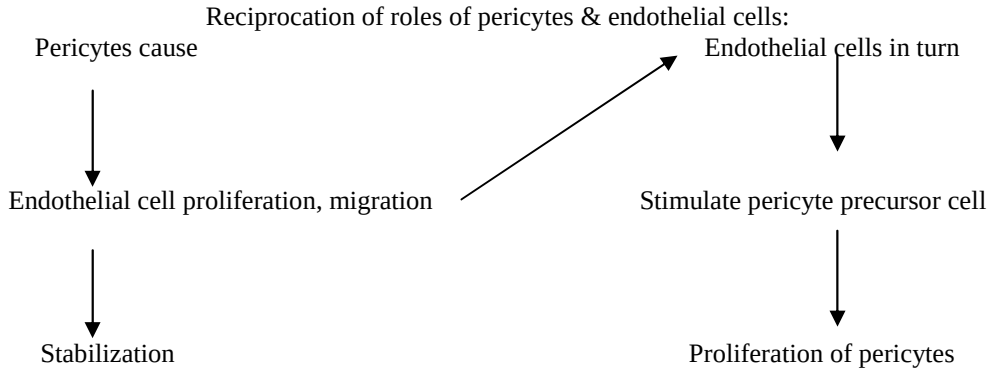


Figure shows hand drawn illustration showing relationship of pericytes to endothelium



Thus, there is a coordination between the number of endothelial cells and pericytes. The balance is highly controlled by the numerous signaling pathways which function in an autocrine and paracrine manner.[2,4] Immunohistochemical analysis is of great help to distinguish between other tumors of the vasculature and wide range of connective tissue tumors from hemangiopericytomas. Molecular markers that are traditionally employed for establishing a definitive diagnosis include CD31, CD34, CD68 alongwith vimentin and also, certain cytokeratins. These markers however are specifically used in diagnosis of connective tissue neoplasms. But these also act as an aid in recognition of mutated stem cells that surround the blood vessels.[5]

Hemangiopericytomas can show varied biological behaviors right from being a slow growing mass to a growth with an aggressive growth having characteristics of a malignancy. Sometimes, its behavior lies in between these two and such type of hemangiopericytoma is known to have borderline or intermediate type of behavior.

Examination of abnormal/atypical mitoses, cellularity, pleomorphism, tendency to bleed and necrosis of the tumor to determine the malignant behavior in a study showed no signs of recurrence or metastasis after 9 months.[5,6] However, in contrast, Enzinger and Smith found that majority of the cases in their study developed recurrences at locations nearby the primary tumor before metastasizing. The most preferable organs for metastasis of this tumor were the lungs. Hemangiopericytomas, specially the malignant ones, are very notorious and in some cases, recurrence and also, metastasis develops after many years of treatment. Thus, it is recommended by a number of studies that patient should be kept essentially under a long-term follow-up even after radical and extensive resection of the tumor.[6,7]

The treatment of hemangiopericytomas lies primarily with wide base resection. Underestimating this entity is risky because of its wide behavior range; thus, a generous resection with extended wide margins becomes unavoidable. Similar procedure was advocated in our case. Adjuvant therapies such as radiation therapy are most often used in cases which are inoperable or which have reappeared after optimal treatment or for palliative care. Chemotherapy however is not routinely employed and its efficacy in the treatment is still not known. [8,9,10]

The differential diagnosis of this lesion includes a broad spectrum of lesions ranging from fibroma, pyogenic granuloma, peripheral giant cell granuloma and fibrous histiocytoma to malignancies such as malignant fibrous histiocytoma and synovial sarcoma. Certain other rarer stromal sarcomas such as fibrosarcomas, mesenchymal chondrosarcomas, vascular leiomyomas and juvenile hemangiomas are also considered in the differential

diagnoses.[11] Fibrous histiocytoma shows a storiform or cartwheel pattern and a less prominent vascular network. Synovial sarcoma may show a biphasic cellular pattern and include fibrosarcoma-like areas. Mesenchymal chondrosarcoma cells are smaller than those of a hemangiopericytoma, and well-defined islands of cartilage are present.[12,13] Reticulin staining can be helpful in such cases to arrive-at a definitive diagnosis of hemangiopericytomas as it is a special stain that demonstrates lesional vessels lined by a single layer of endothelial cells with the pericytes covering the basement membrane of blood vessel (Fig.7).[13,14] Histopathology of hemangiopericytomas plays a crucial role as the treatment of this lesion is dependent on the amount of cellular atypia and mitotic activity present in the lesion. The more bland lesions with minimal mitotic activity are treated by a wide base excision while the more active and dysplastic lesions are treated by radical excisions with or without adjunctive radio and chemo-therapies.[14,15]

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