CASE REPORT

Hemangiopericytoma of mandible

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ABSTRACTS
Hemangiopericytoma is a rare vascular neoplasm, which arises from specialized cells (pericytes) around the capillary walls. Only 5% of hemangiopericytomas in the series of Stout occurred in the oral cavity and pharynx. Several studies have revealed that due to chances of late recurrence or metastasis, long-term follow-up is necessary in patients with this tumor even after radical resection. We report a case of hemangiopericytoma of mandible in a 26-year-old woman, with 4 years' follow-up.
Key words: Hemangiopericytoma mandible, vascular neoplasm

INTRODUCTION

Hemangiopericytoma is a very rare vascular neoplasm characterized by the proliferation of capillaries surrounded by masses of round or spindle-shaped cells.[1] Stout and Murray first described this tumor in 1942.[2] The tumor has a predilection for the long bones, pelvis, and scapula, but 15% occur in the head and neck region. Origin in oral cavity is less common. We report a case of hemangiopericytoma of the mandible in a 26-year-old woman.

CASE HISTORY

A 26-year-old woman reported with complaint of swelling at the right body of the mandible of 1-year duration. History revealed that a year ago, her right last lower molar was extracted as it had become mobile and tender. After few days of the extraction, she noticed development of the swelling in the region. There was no history of any trauma, pus discharge, or fowl smell from the site. Local examination revealed diffuse swelling, seen on the right body of the mandible extending from right lower first molar to ascending ramus of the mandible. Overlying mucosa appeared normal in color. On palpation, the swelling was hard, nontender, nonfluctuant, and approximately $3 \times 2 \times 1.5$ cm in size. Both buccal and lingual cortical plates were expanded. Oral hygiene of the patient was satisfactory. All the teeth were periodontically sound. No engorged vessels or discharging sinus was noted. No regional lymphadenopathy was found.

Panorex x-ray showed a multilocular radiolucent lesion in the right body and ramus of mandible, involving whole of the coronoid process [Figure 1]. CT scan mandible showed expansile lytic lesion in the right body, ramus, and coronoid process of mandible with break of cortex medially at the molar region.

Incisional biopsy was taken under local anesthesia and diagnosed histologically as hemangiopericytoma of mandible. Biopsy revealed cellular neoplasm with variably dilated blood vessel under low magnification (H and E, ×40)

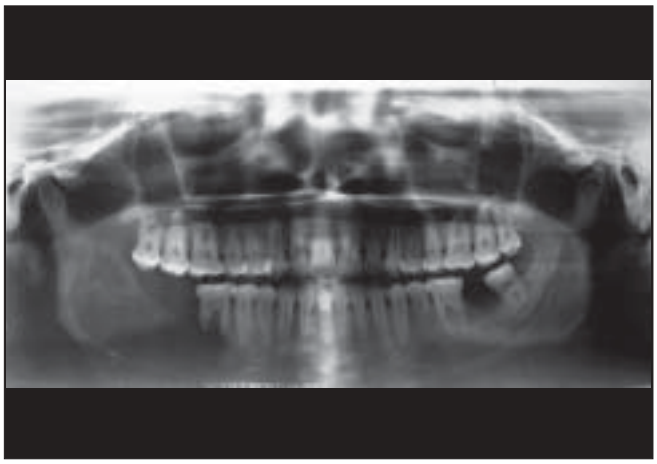

Figure 1: Showing multilocular radiolucency involving right body and ramus of the mandible

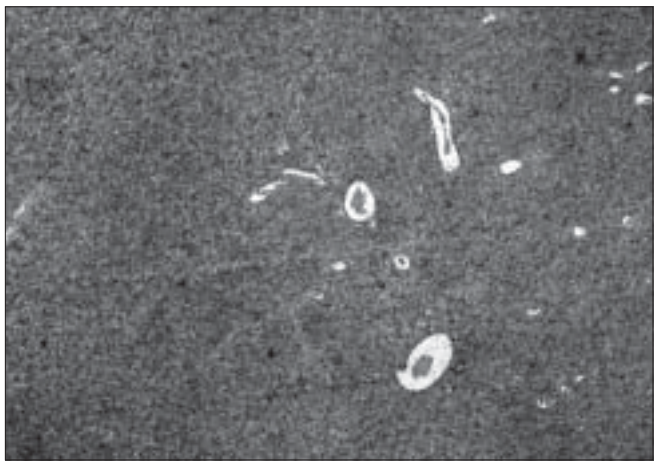

Figure 2: Photomicrograph revealing cellular neoplasm with variably dilated blood vessel (H and E, 40×)

[Figure 2]. High magnification (H and E, ×400) revealed cells with ill-defined cell margin, round-to-oval nuclei with granular chromatin but without any significant mitosis or any areas of necrosis [Figures 2 and 3]; and reticulin stain revealed the vessel wall with the tumor cells to the outside of the highlighted branching vessel wall [Figure 4]. Skeletal survey was negative. Hemi-mandibulectomy on the right side sparing the condyle [Figure 5] was undertaken under general anesthesia, and reconstruction of mandible was done with titanium reconstruction plate and screws [Figure 6]. The patient was discharged after an uneventful recovery. Patient is on regular follow-up since last 4 years. No signs of recurrence or metastasis have been noted so far.

DISCUSSION

Hemangiopericytomas have been described in all age groups; with more than 40% occurring in the fifth and sixth decades.[3] This tumor has no sex predilection. Clinically the lesion is firm, apparently circumscribed and often nodular, and may or may not exhibit redness indicative of their vascular nature. A painless enlarging mass is the general mode of presentation, as seen in this case. Majority of tumors grow rapidly and are therefore of short duration.[4]

The radiographic feature of a hemangiopericytoma is that of a malignant bone lesion but is nonspecific. Histologically, the tumor is characterized by tightly packed cellular areas surrounding thin-walled branching blood vessels. The tumor cells are small, ovoid to spindle shaped with ill-defined cell boundaries. The collapsed blood vessels lined by flat endothelial cells are arranged in a 'stag-horn' pattern. The spindle cells are each surrounded by a reticulin sheath and are separated from the endothelial cells by a basement membrane. Silver stain demonstrates extravascular location of the tumor cells, and individual cells display pericellular reticulin pattern.[5] The blood vessels are lined by normal endothelium in contrast to malignant angiosarcoma, where the vascular spaces are lined by malignant tumor. Histologically, several other tumors may show a vascular pattern, resembling hemangiopericytoma. Fibrous histocytoma, synovial sarcoma, and mesenchymal chondrosarcoma should be considered in the microscopic differential diagnosis. Fibrous histocytoma shows a storiform or cartwheel pattern, and a less prominent

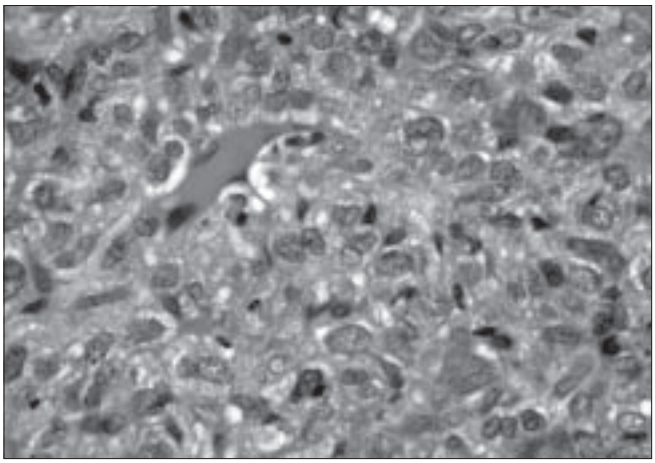

Figure 3: Photomicrograph showing monophasic cells around blood vessels (H and E, 400×)

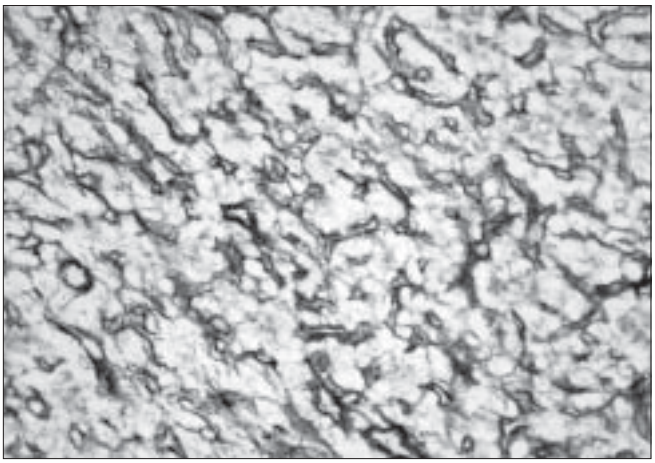

Figure 4: Reticulin stain highlights the extravascular presence of monophasic tumor cells

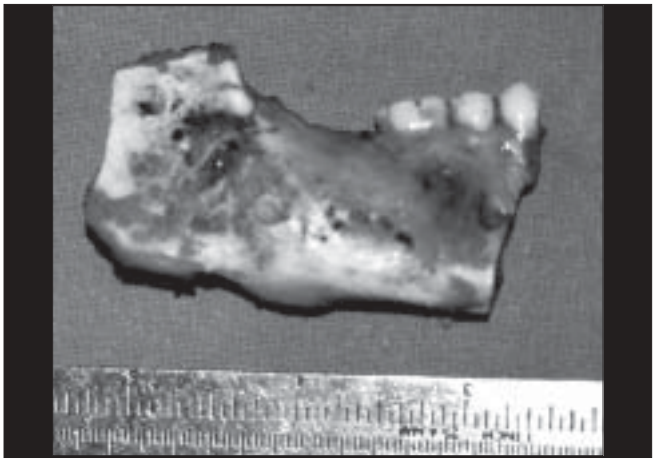

Figure 5: Tumor specimen

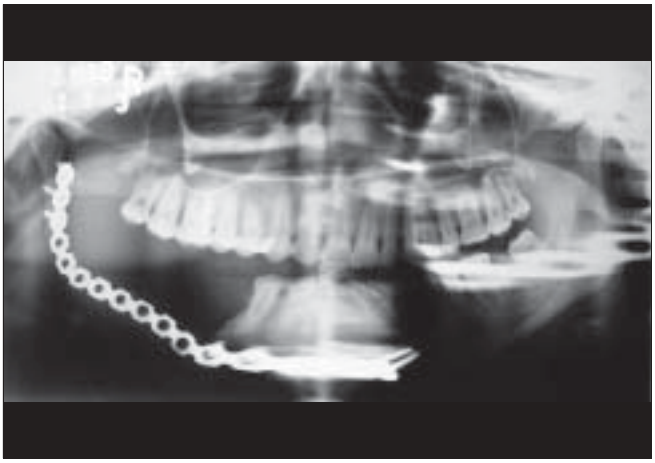

Figure 6: Postoperative radiograph

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.[1]

The treatment of hemangiopericytoma of bone is primarily surgical. Initial wide resection is mandatory if a cure is to be expected, which was done in the present case. The role of chemotherapy is unknown. Radiation therapy has usually been reserved for unresectable and recurrent tumors or the achievement of palliation.[6]

Enzinger and Smith[5] found that in more than two thirds of the cases that eventually metastasize, develop local recurrences before metastasis. Lungs are the most common sites of metastasis. Several studies have shown that long-term follow-up is necessary in patients even after radical resection because recurrence or metastasis may be delayed by many years.

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