Hemangiopericytoma of the Maxilla in a Pediatric Patient: A Case Report

Rohit Anand, MDS *Sonal Gupta, MDS*

ABSTRACT

The Hemangiopericytoma is a malignant vascular tumor arising from mesenchymal cells with pericytic differentiation. Hemangiopericytoma is most commonly seen in adults, and only 5% to 10% of cases occur in children. The tumor is extremely rare in the head and neck region (16%)[1]. Cytogenic abnormalities have been present in some hemangiopericytoma cases. Surgical resection remains the mainstay treatment. Adjuvant chemotherapy and radiotherapy is appropriate for cases of incomplete resections and life-threatening tumors particularly in children. Late relapses may occur and require long-term follow-up. A 4-year-old child patient with hemangiopericytoma of the maxilla presented with firm, recurrent, but painless jaw mass. Radiographic investigations revealed a poorly circumscribed radiolucency. The lesion biopsy showed well-circumscribed multiple lobules of tumor mass consisting of tightly packed, spindle-shaped cells. Chemotherapy and radiotherapy of the lesion was conducted. The role of the pediatric dental team is extensive in children with hemangiopericytoma, who require a regular dental review. The patient's oncologist should be immediately contacted if there is any suspicion of recurrence.

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Hemangiopericytoma is a very rare, slow-growing vascular tumor with variable malignant potential, constituting less than 1% of all neoplasm. The tumor is extremely rare in the head and neck region.

The first description of this tumor was provided by Stout and Murray[1] in 1942. The tumor takes its origin from pericapillary pericytic cells. On histopathological examination, the tumor is a characterized by a proliferation of oval and spindle-shaped pericytic cells. On first diagnosis, it is generally difficult to distinguish between benign and malignant forms. This frequently becomes evident only as the disease progresses. The features of malignant transformation are increased mitotic activity, higher cell density, appearance of undifferentiated cells, and necrotic and hemorrhagic zones in the tumor tissue. Cytogenic abnormalities have been found in hemangiopericytoma. Hallen et al[2] concluded that most hemangiopericytoma are near diploid, and break points in 12q13, 12 q 24, and 19q13 seem to be common, with recurrent *t* (12; 19; q 13; q 13) translocation.

Children with this lesion are likely to present to the pediatric dental department, as illustrated in the following case. The purpose of this report was to describe the presentation, investigation, diagnosis, and treatment of hemangiopericytoma in the maxilla of a pediatric patient.

CASE REPORT

A 4-year-old male child presented to the department with a firm, diffuse swelling of the maxilla on his left side that had been present for 3 months . The swelling was painless. This had not improved with antibiotics prescribed by a general dental practitioner.

Clinically, the boy was pale, with nonpalpable, nontender lymph nodes in the submandibular region. Extraoral examination showed a single diffuse swelling of 3x4 cm in size on his face's left side involving the alveolar part of the maxilla. The margins of swelling were ill

Dr. Anand is senior lecturer in the Department of Pedodontics, Sardar Patel Post Graduate Institute of Dental Sciences, Lucknow (UP), India. Dr. Gupta is residing at Agra, India.
Correspond with Dr. Anand at rohitanand555@yahoo.com

defined. There was obliteration of the nasolabial fold, and the left ala of his nose was deviated. The philtrum was slightly deviated to the right side. The skin over the swelling was normal, and there was no surface pulsation.

Intraorally, growth extended both labially and palatally in regions 61 and 64. On the labial side, lesion extended superiorly up to the labial sulcus and inferiorly 3 mm below the occlusal line. The palatal aspect showed indentation of mandibular teeth observed on the swelling (Figure 1). There was displacement of upper left primary canine. The lesion showed bleeding on provocation, consistency was firm, and growth was sessile.

PROVISIONAL DIAGNOSIS

The provisional diagnosis of the anterior maxilla's neoplasm revealed the following tumors: fibrous sarcoma; hemangiopericytoma; chondrosarcoma; Burkitt's lymphoma; malignant peripheral nerve sheath tumor; thymomas; and osteosarcoma.

A provisional diagnosis of hemangiopericytoma was made, and relevant hematological, radiological, and biochemical investigations were undertaken to confirm the aforementioned.

RADIOGRAPHIC INVESTIGATIONS

Occlusal radiograph revealed a poorly circumscribed radiolucency involving upper left primary and permanent teeth. A computed tomography scan showed an osteolytic lesion involving the maxillary left alveolar ridge and soft tissue attenuation measuring up to 1.2x1 cm (Figure 2).

BIOPSY

Incisional biopsy of the lesion showed well-circumscribed, multiple lobules of tumor mass consisting of tightly packed, spindle-shaped cells around ramifying, walled, endothelium-lined vascular channels showing a stag horn configuration (Figure 3). The cells have round to oval nuclei with a moderate amount of cytoplasm. There are district intravascular and perivascular satellite nodules outside the main tumor mass. There is evidence of intravasular endothelial cell proliferation. Mitotic cells are together with focal necrosis and vascular invasion.

IMMUNOHISTOCHEMISTRY

Actin showed a positive expression with a diffuse pattern to the tumor cells (Figure 4). Vimentin also showed a positive expression to the tumor cells.

FINAL DIAGNOSIS

The various investigations shows that the final diagnosis of the lesion is hemangiopericytoma of the anterior maxilla.

MANAGEMENT

Chemotherapy and radiotherapy of the lesion was performed. No perforation of the cortical plate was noted at the time of treatment. Currently, the patient remains healthy and is under regular review.

DISCUSSION

Hemangiopericytoma is a soft tissue tumor derived from mesenchymal cells with pericytic differentiation. It can either be benign or malignant. Two subtypes have been described: (1) infantile hemangiopericytoma in infants younger than 1-year-old; and (2) adult hemangiopericytoma in anyone older. The pediatric cases account for approximately 3% of all soft tissue sarcomas in this age group. Most infantile hemangiopericytomas are considered congenital; they represent approximately one third of all pediatric hemangiopericytomas.[3]

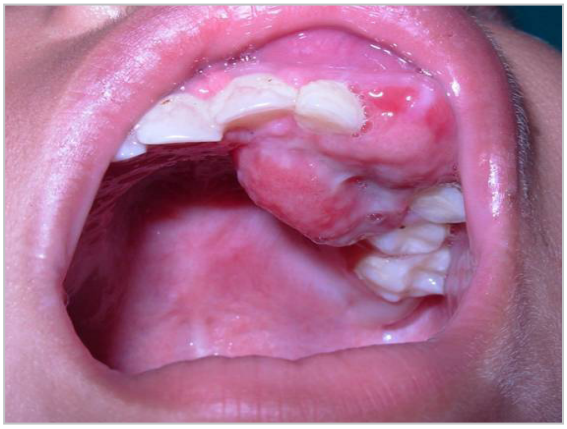

Figure 1. Intraoral view of the case showing a lesion labially and indentations of mandibular teeth seen on the swelling.

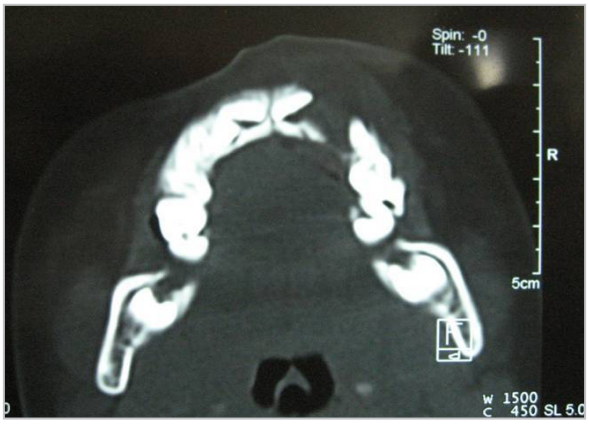

Figure 2. Computed tomography scan showing an osteolytic lesion involving the maxillary left alveolar ridge and soft tissue attenuation.

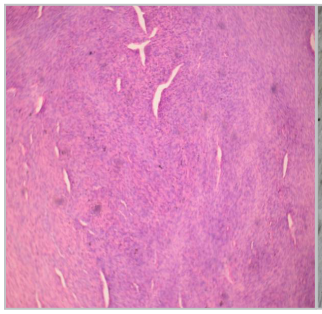

Figure 3. Clinical micrograph showing multiple lobules of tumor mass consisting of spindle-shaped cells.

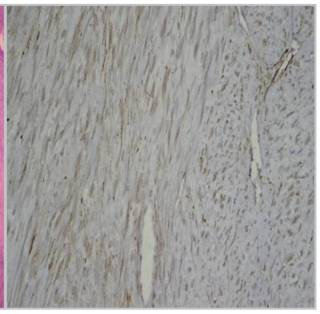

Figure 4. Clinical micrograph revealed that Actin showed a positive expression with a diffuse pattern to the tumor cell.

The tumor may occur anywhere in the body, with the most common anatomic locations being the lower extremities, the pelvis, and the head and neck region.[4] Hemangiopericytomas are usually deep seated and found in the muscle tissue; dermal and subcutaneous hemangiopericytomas are much less common. Clinical presentation of a hemangiopericytoma's nonspecific pain is a late symptom associated with an enlarging mass.[5]

Hemangiopericytoma in children is very rare, so only few reports or data about its clinical management have been published. Surgical resection is the main treatment.[6] Child hemangiopericytomas are characterized by high response to chemotherapy, which is required when tumors are life-threatening.[7] Positive responses to chemotherapy have been reported with vincristine, cyclophosphamide, deoxorubicin, dactinomycin, methotrexate, mitoxantrone, and other alkylating agents. Considering the high chemoresponsiveness of tumors as well as the early age of patients, less toxic chemotherapeutic regimens that limit use of authracyline drugs and/or alkylating agents need to investigated.[8] Radiotherapy is effective in some patients, but indications are limited because of a child patient's age.

A hemangiopericytoma's prognosis is usually favorable, depending on the tumor's mitotic activity. The number of mitotic figures are variable and of prognostic significance. Espat et al[9] reported that 2- and 5-year overall survival rates were 93% and 86%, respectively. They also reported that incidence of metastases varies from approximately 10% to 60%, depending on the diagnostic criteria and the therapy. Local and distant relapses after a prolonged, disease-free interval have been reported by Sptiz et al,[10] suggesting a mandatory long-term follow-up.

CONCLUSIONS

In hemangiopericytoma, child patient prognosis and long-term survival can vary greatly from individual to individual. Prompt medical attention and aggressive therapy are important for the best prognosis. Continuous follow-up care by the pediatric dental team is essential for a child with hemangiopericytoma. Radiation and chemotherapy side effects and tumor recurrence can occur in hemangiopericytoma survivors.

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