CASE REPORT

A Rare Case of Hemangiopericytoma in the Maxilla of a 4-Year-Old Child

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ABSTRACT

Hemangiopericytoma is a rare vascular tumor that originates from small pericapillary spindle-shaped cells. Clinical diagnosis of such a lesion is an extremely challenging task because there are very few detailed reports of such a lesion and one would hardly think of a diagnosis such as hemangiopericytoma while examining a lesion. The incidence in head and neck region is less than 20%. Hemangiopericytoma usually occurs in the 4th to 5th decade of life and just 3 to 5% cases affecting the soft tissues around the oral cavity, sinus tract and meninges. The hemangiopericytoma generally presents as a painless mass, but may have symptoms depending upon the size and location of the tumor. Here, we present such a case in a 4-year-old female child involving the anterior maxillary region.

Keywords: Hemangiopericytoma, Vascular neoplasm, Staghorn, Immunohistochemistry.

INTRODUCTION

Hemangiopericytoma is an uncommon mesenchymal neoplasm, first described by Stout and Murray in 1942. Hemangiopericytoma is considered to be a tumor of pericytes. Apart from being rare in absolute terms, hemangiopericytoma is uncommon in the head and neck. Most of the cases described till now are usually in the trunk or lower extremities.1 The incidence in head and neck region is less than 20%. In this site, it affects soft tissues around the oral cavity, sinus tract and meninges.2 In our case, the tumor involved the anterior maxillary region. The hemangiopericytoma generally presents as a painless mass, but may have symptoms depending upon the size and location of the tumor. In the discussion, the lesion had similar features but also caused difficulty on eating and talking.3

CASE HISTORY

A 4-year-old female patient visited to the department of oral medicine and radiology with the chief complaint of swelling in the upper left front region of the jaw since three months, which started as a slight small and rapidly increased in size, and attained the present size not associated with any pain or fever. The past medical and dental histories were non significant as well as personal history and family histories were noncontributory.

General physical examination, gait was normal and moderately built and nourished. All vital signs were within normal range. Extraoral examination revealed a single, diffuse swelling on the left side of the face measuring 3 × 4 cm extending anteriorly from the philtrum and posteriorly to the left commissure of the mouth. Margins of the swelling were diffuse, surface was smooth, nasolabial fold was obliterated and the upper lip was slightly elevated. No secondary changes and no visible pulsations seen. No change in color was noticed.

On palpation, no localized raise of temperature, firm in consistency and no tenderness. Lymph nodes were not palpable. Intraoral examination revealed a proliferative, sessile growth seen on the left front region of maxillary alveolar mucosa measuring 3 × 4 cm irt 61, 62, 63 with ill-defined margins. The growth extended labiopalatally with extrusion of 62 and displacement of 63 palatally. Indentations of mandibular teeth were seen on the growth. Overlying mucosa was pinkish red with no discharge or surface pulsations. Bleeding on probing was present on palpation and the growth was fixed to the underlying bone (Figs 1 and 2).

On the basis of history and clinical examination, a provisional diagnosis of malignant neoplasm of anterior maxilla was given with differential diagnosis of juvenile ossifying fibroma, fibrosarcoma, Burkitt’s lymphoma, Ewing’s sarcoma, chondrosarcoma and hemangiendothelioma.

Occlusal maxillary topographic radiograph revealed erupting permanent tooth buds with cortical bone destruction and extrusion of 62.

CT revealed osteolytic lesion involving left maxillary alveolar ridge with soft tissue attenuation measuring upto 1.2 × 1 cm; no evidence of sclerosis or any calcific density (Fig. 3).

Histologically, well-circumscribed multiple lobules of tumor mass consisting of tightly packed spindle-shaped cells around ramifying-walled endothelium lined vascular channels showing a staghorn configuration. The cells have round to oval nuclei with moderate amount of cytoplasm. Reticulin stains reveal proliferating neoplastic cells outside the vessels. Dense reticulin meshwork surrounding vessels and tumor cells were seen (Fig. 4).
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In immunohistochemistry, Actin showed positive expression with a diffuse pattern and Vimentin was also positive (Fig. 5).

Management was done by surgical excision of the lesion.

DISCUSSION

Hemangiopericytoma mostly arises in the 4th to 6th decade of life and is rarely seen in children. Pediatric cases account for less than 10% of all hemangiopericytoma. The etiology is clearly known, although the presence of hemangiopericytoma has been linked to trauma, prolonged steroid use and hormonal imbalance. In our case, the patient gave no such history. The lesion had arisen de novo.

Diagnosis of hemangiopericytoma is a challenging task because it is rare in occurrence and has no specific clinical behavior. It
needs to be differentiated from chondrosarcoma, Ewing’s sarcoma and osteosarcoma, Juvenile hemangioma and leiomyosarcoma. In our case, the age group, rapidity of growth and other clinical features favored a provisional diagnosis of sarcoma.5

Radiographically, the lesion may show destruction of bone or well circumscribed, soft tissue mass that often displaces neighboring structures. Under light microscopy, hemangiopericytoma is characterized by vascular channels arranged in staghorn manner.6 These vascular spaces are interconnecting in a ramifying configuration, compressed or even obliterated and the tumor cells are arranged in compact sheets around the thin-walled vascular channels.7 Immunohistochemically, the hemangiopericytoma cells are invariably positive for Vimentin but very rarely and only focally for smooth muscles markers such as Actin and desmin.8

Histopathologically, all the features were present in our case. The management of hemangiopericytoma involves wide surgical excision. In the head and neck, cervical lymphadenectomy is reserved for those instances, where palpable adenopathy is coexistent. The role of radiotherapy has been questioned.4

The present patient underwent wide excision. Radiotherapy was not advised as there was no lymphadenopathy. The chances of it to recur locally after years or even decades following initial treatment have been reported and the patient should be followed up for the lifetime with a minimum period of atleast 10 years. The patient is kept under observation and regular follow-up, and remains free of recurrence since three years.

In conclusion, although rare, hemangiopericytoma should be considered as one of the differential diagnoses of tumors of the orofacial region. It continues to be a very diagnostic and therapeutic challenge, hence every diagnosed case of hemangiopericytoma could be a contributing factor.

REFERENCES