

Hemangiopericytoma- A case report

¹Dr Nitin Agarwal, Prof & head, Dept of OMDR, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow, U.P, India.

²Dr. Nisha Kumari, PG student, Dept of OMDR, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow, U.P, India.

³Dr K.k. Chaudhary, Assistant Professor, Dept of OMDR, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow, U.P, India.

⁴Dr Somi Fatima, Assistant Professor, Dept of OMDR, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow, U.P, India.

⁵Dr Anchal Rai, PG Student, Dept of OMDR, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow, U.P, India.

⁶Dr Nadia Irshad, PG student, Dept of OMDR, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow, U.P, India.

Corresponding Author: Dr. Nisha Kumari, PG student, Dept of OMDR, Career Post Graduate Institute of Dental Sciences and Hospital, Lucknow, U.P, India.

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Abstract

Hemangiopericytoma is a rare neoplasm of pericytes forming the wall of capillaries. It is mostly seen in 3rd to 5th decade of life, rarely occurs in young age. These tumors can originate anywhere in the body where there are capillaries. The most common locations reported are the brain, lower extremities and pelvic area, incidence in head and neck is comparatively low. It can either be benign (non-cancerous) or malignant (cancerous). Malignant tumors can metastasize to different parts of body primarily lungs and bones. These tumors are painless masses, often

associated with no symptoms. They can remain undetected for longer period of time, due to the fact that these tumors arise in soft tissues which create room for the new mass. When occurs in brain, can mimic meningiomas causing neurologic disturbances. Hereby, we report a rare case study of Hemangiopericytoma of mandible in a 6 year old patient. Thus this study helps to evaluate not all the soft tissue growth occurring in oral cavities are pyogenic granuloma, peripheral giant cell granuloma and fibroma but it can be a rare case of Hemangiopericytoma too.

Keywords: Benign, Malignant, Hemangiopericytoma, Rare, Pericytes, Metastatize

Introduction

Stout in 1942 was the first to report an oral hemangiopericytoma. It is a distinct vascular neoplasm which is usually benign but has a definite malignant counterpart. Mostly seen in 3rd to 5th decade of life, occurrence before 2nd decade is rare with no sex predilection. Accounts for 1% of all blood vessel-related neoplasm and 2-3% of all soft tissue sarcomas. Hemangiopericytoma is a soft tissue sarcoma that originates in the pericytes forming the wall of capillaries. Pericytes are modified smooth muscle cells present outside the reticulin sheath of the endothelium. Hemangiopericytoma exhibits an unforeseeable biologic behaviour, when diagnosed they are often low-grade benign sarcomas which commonly represent slow growing and painless tumors of the extremities. Malignant forms show necrosis, cellular pleomorphism, high proliferative index and mitoses >4 per 10 high power fields. The absence of necrosis, cellular pleomorphism, mitoses <4 per 10 high power fields does not necessarily indicate benign nature. In fact tumour with benign histological appearance has been reported to metastasize. Patient with a past history of trauma, prolonged steroid use and hypertension have shown the prevalence of these tumours although no fixed etiological factor is identified. Chromosomal translocation t(12:19) and t(13:22) have been observed in the tumor. Tumor cells are immunoreactive for vimentin, HLA-DR antigen and CD-34. As sarcomas, hemangiopericytomas are graded on the basis of histological and biological parameters. However, as the pericytes possess characteristics of both smooth muscle cells and endothelial cells, differentiating these from other cell types becomes challenging. Accordingly, diagnosis is made based on architectural patterns

exhibited histologically. The disease tends to recur after surgical excision, making it mandatory to follow-up for longer period of time.

Case report

A 6 years old male patient reported to the department of oral medicine and radiology with a chief complaint of growth in lower left back region of the jaw since 5-6 months, which was gradually increasing in size. A detailed base of the mandible supero-inferiorly and commissure of the lip to 1 cm below tragus of the ear antero-posteriorly measuring up to approx. 4x2 cm in size was seen. The swelling was firm and non-tender on palpation. There was no distant or regional lymphadenopathy. Patient underwent surgery at the same site 1 year ago and came back again for the recurrence of the same after 5 months of the previous surgery.

On intraoral examination, a well-circumscribed lobular growth was seen on the left side of the jaw involving the distal surface of the first molar measuring upto 2.5x1.5 cm in size. It was sessile, firm and non-tender on palpation. The mass was causing obliteration of the left vestibular mucosa.

After the routine haematological investigations, which were almost within the normal limits, the patient was subjected to radiographic evaluation. Orthopantomogram didn't reveal any significant finding, single multilobular radiolucency seen overlying the bony crypt of erupting second molar without any evident bony invasion. Based on the above clinical features and radiological features a provisional diagnosis of Peripheral giant cell granuloma and a differential diagnosis of fibroma was made. After which an excisional biopsy was performed that showed spindle shaped cells with fibrous tissue and small blood vessels confirming the diagnosis of hemangiopericytoma.

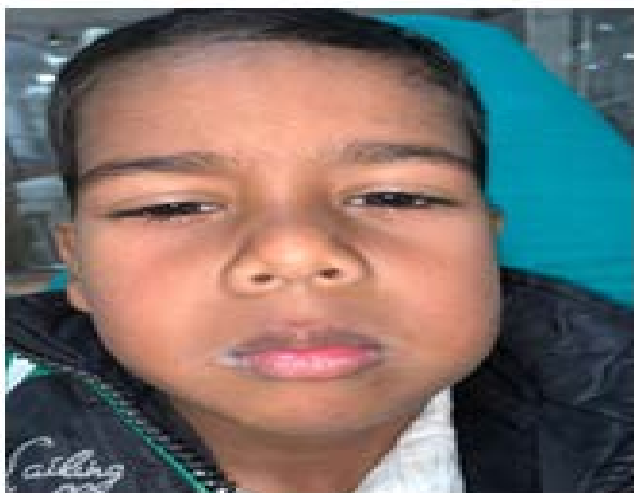


Fig 1: Preoperative clinical photograph of patient with swelling on the left side of the face



Fig 4: Post-operative clinical photograph of the patient



Fig 2: Showing intraoral growth on the left side of the mandible



Fig 3: Orthopantomograph of the patient shows no invasion of bone

Discussion

Hemangiopericytoma was defined in 1942 by Stout and Murray as a tumor that arises from pericytes surrounding capillary vessels. It can occur anywhere in the body where capillaries are present. Most common location in body is in the limbs and retroperitoneum while it is rarely seen in head and neck region, if present mostly occurs in buccal mucosa, lips, maxilla and mandible (1,2).

Hemangiopericytoma accounts for less than 1% of all vascular tumours. Many pathologists suggested that Solitary fibrous tumor and Hemangiopericytoma are two spectrums of same disease based on histological features. However, Hemangiopericytoma has been included in extra-pleural Solitary fibrous tumor. The cellular end of this spectrum corresponds to classic Hemangiopericytoma whereas the hyalinised end referred to Solitary fibrous tumor(4). According to WHO 2002 classification, it is neither classified as benign nor malignant rather a benign lesion with low grade malignancy(1). The etiology remains unknown, although some factors like previous history of trauma has raised the concern that this may stimulate proliferation of pericytes following damage to the wall of capillaries, long term steroid therapy, and arterial hypertension have been implicated in causing

Hemangiopericytoma, but none of the theories have been proved. Cytogenic abnormalities are being studied and most Hemangiopericytomas are near diploid, and break points in 12q13, 12q24 and 19q13 are common with recurrent t(12;19)(q13;q13) translocation(5). A firm, solitary painless enlarging mass with local infiltration is the general mode of presentation as in this case (3, 5). These tumors have tendency to metastasize early or late through vascular and lymphatic route. Rate of metastasis of malignant variety is reported to be less than 75%. A rare case of metastatic hemangiopericytoma to lung after 20 years of its initial occurrence in mandible was reported by Ravenel and Goodman which emphasis.

Conclusion

Wide surgical excision is the most accepted treatment for Hemangiopericytoma, although surgical resection by itself poses a challenge for the surgical team because of the size, extent and proximity of the lesion to vital anatomical structures. Early diagnosis and management of these lesions can limit post-surgical morbidity. Since local recurrences are very common and distant metastasis has been reported to occur even after 20 years after primary tumor, long term follow-up becomes essential both clinically and radiographically.

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