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CASE REPORT

Hemangiopericytoma of Gingiva in a 4-Year-Old Child: A Rare Case Report

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Abstract A hemangiopericytoma is a type of soft tissue sarcoma that originates in the pericytes in the walls of capillaries which was characterized in 1942. It is commonly seen in the age group of 5th–6th decades of adults and only 5–10% of cases occur in children with extreme rare occurrence in head and neck region (16%). A rare case of hemangiopericytoma in a 4-year-old female patient is presented here and its clinical, histopathological and immunohistochemically features are discussed. Though surgical resection remains the mainstay, excisional biopsy was primarily performed to reach the final diagnosis. Even in the follow-up phase of 1 year, no recurrence or no metastatic changes were observed.

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Introduction

Hemangiopericytoma/solitary fibrous tumour is a very rare slow-growing vascular tumour of uncertain malignant potential constituting less than 1% of all neoplasm [1]. The first description of this tumour goes back to Stout and Murray [2]. Under the WHO classification [3], hemangiopericytomas and solitary fibrous tumours of the soft tissues are regarded as features of same entity in soft tissue fascicle. It originates in pericytes which are the cells of mesenchymal origin and partially surround the endothelial cells of capillaries and veins. There is still a controversy over the histogenesis and histological classification of hemangiopericytoma. It is no longer considered a specific entity, but instead a growth pattern shared by a variety of unrelated soft tissue neoplasms and should be considered a diagnosis of exclusion [4].

It is most commonly seen in 5th–6th decades of life with only 5–10% cases occurring in childhood [5]. Fortunately, it is rare and typically benign. It occurs most commonly in the soft tissues of upper and lower extremities and retroperitoneum. Incidence of HPC is uncommon (16%), and lesion arising from the gingiva has rarely been reported. In a meta-analysis by Brockbank from 1949 to 1979, only 35 cases were reported in oral cavity including—Tongue (9), upper jaw bone (5), lips (4), buccal region (3), cheeks (3), gingiva (2), parotid gland (2) and multifocal lesions [6].

The diagnosis of hemangiopericytoma is made based on its characteristic architectural pattern and configuration in ultrastructural or immunohistochemical studies [7].

As only a few cases have been reported in the literature, we present a case report to describe the presentation, investigation, diagnosis and treatment planning of hemangiopericytoma in both the arches originating from gingiva of a paediatric patient.

Case Report

A 4-year-old female patient presented with a painless growing mass from gingival tissue in left mandibular and bilateral maxillary molar teeth area (palatally) for last 7–9 months.

Past History of Trauma and Present Medical History—Patient had a history of trauma to left side of head due to fall from terrace 1 year back. After trauma, she was unconscious for 3–4 min and even had a history of epileptic attacks since then, though clinical findings could not be correlated to any radiological findings on CT and MRI. Since then, she had been under medication for epileptic attacks (syrup sodium valproate and levetiracetam).

Extra oral Examination presented with bilaterally symmetrical face with no abnormality detected. On intra oral examination, growth extended on the palatal aspect of both primary maxillary molars (approaching each other in the mid-palatal region) (Fig. 1) and on buccal aspect of left lower primary molars (Fig. 2). It was pinkish red in colour, lobulated and measured approximately 4×3 cm^2 in maxilla bilaterally and 1×1.5 cm^2 in mandible. Base of lesion was pedunculated and did not bleed profusely on bleeding and consistency was firm. There was no mobility observed in teeth associated with the lesion in both maxilla and mandible. Oral hygiene of the patient was poor.

Provisional Diagnosis

The provisional diagnosis of neoplasm in maxilla are following tumours—fibrous sarcoma, hemangiopericytoma, chondrosarcoma, Burkitt's lymphoma, malignant peripheral nerve sheath tumour, thymomas and osteosarcoma.

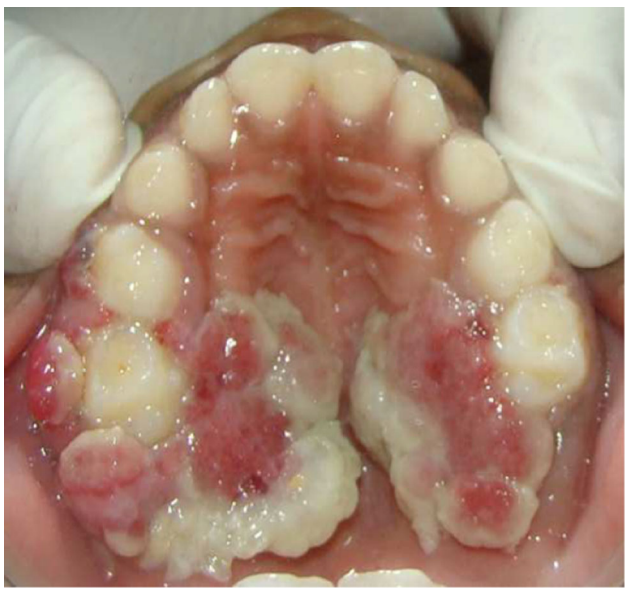

Fig. 1 Extension of palatal growth in maxilla

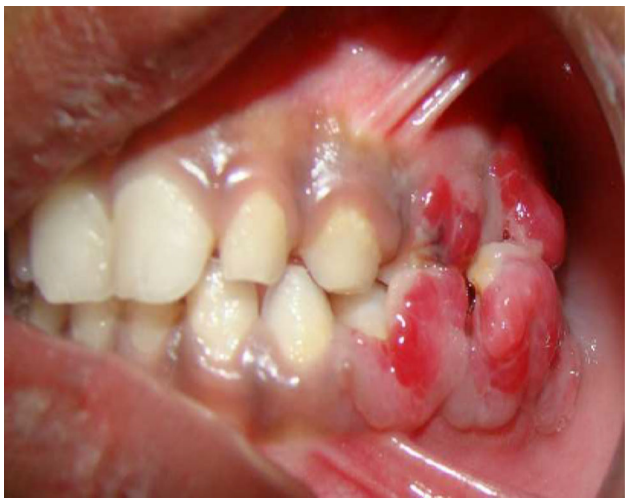

Fig. 2 Extension of growth in mandible

As per the present medical history, patient was under medication sodium valproate and levetiracetam for epileptic attacks, a differential diagnosis of drug induced gingival hyperplasia was also considered.

Biopsy

Under general anaesthesia, excisional biopsy of the lesion was performed using monopolar electrocautery to obtain good haemostasis and avoid the need of sutures. There was no other surgical intervention needed, subsequent to excisional biopsy.

On histopathological examination, soft tissue section showed a highly cellular connective tissue stroma which consisted of numerous atypical cells. The spindle cells were arranged in a lobular pattern at places and at other areas in irregular fashion. The tumour cells exhibited the presence of various dysplastic features such as a low degree of cellular and nuclear pleomorphism, hyperchromatic and even vesiculated nuclei and few mitotic figures. The tissue was highly vascular with numerous, engorged immature blood vessels of varying size and shape. The blood vessels exhibited a 'staghorn' pattern, presence of a small, round indistinct lumen and were also present in the form of slit-like spaces and the reticulin positivity was observed. Spindle-shaped and plumped tumour cell lining the blood vessels were also filling the stroma. There was scanty amount of chronic inflammatory infiltrate dispersed throughout the tissue, and the connective tissue septae with dense collagenous fibres were present. Overlying epithelium was atrophic to hyperplastic, parakeratinized stratified epithelium with elongated rete ridges at places (Fig. 3).

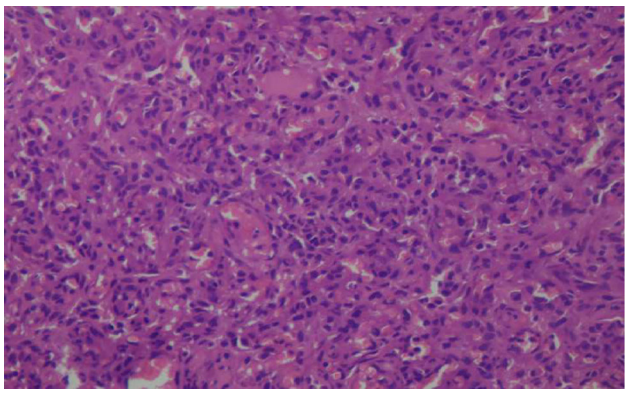

Fig. 3 H&E, ×10 view showed sheets and glomeruli of tumour cells with intervening fibrous connective tissue

Immunohistochemistry

Actin showed positive expression with diffuse pattern by tumour cells. Vimentin was also positive.

Based on these findings, a confirmatory diagnosis of hemangiopericytoma of gingiva was made. As no radiological findings could be correlated to clinical findings, only excisional biopsy of the lesion was performed. Currently, the patient remains well and is under regular review.

Discussion

Stout and Murray first time reported nine cases of tumours derived from vascular pericytes [2].

Hemangiopericytoma, a soft tissue tumour derived from mesenchymal cells with pericytic differentiation, can occur anywhere in the body with the most common anatomic locations being the lower extremities, the pelvis and rarely in head and neck region [8]. About 15–25% of all HPCs occur in head and neck region specially in the nasal cavity, paranasal sinuses, orbital region, parotid gland and the neck and rarely occur in the pharynx [9]. HPC of gingiva in the oral cavity is extremely rare, and its malignant potential is varied. In 2002 WHO classification, it was categorized as neither benign nor malignant, but was regarded as tumour with potential of low malignancy.

HPC commonly presents as a slowly enlarging, painless mass. Pain usually occurs only in large lesions that are locally invasive or confused in yielding spaces such as paranasal sinuses. However, pain is characteristic of bone or pulmonary metastases [10].

In a study by Michi et al. [11], the most common intraoral site of origin was found to be palate followed by mandible, lower lip and most commonly associated symptoms comprised of a mass/swelling ranging from 3 to 60 mm. Out of 16 cases of intraoral HPCs, 8 were described as malignant. Also in an analysis of 106 cases of HPC by Enzinger and Smith [14], there was no difference in sex predilection and the median age was 45 years with 86% of the patients between 20 and 69 years of age. The present case was a female patient of 4 years of age with enlarging mass of 4 × 3 cm^2 in maxilla (present palatally) bilaterally and 1 × 1.5 cm^2 in mandible which was first noticed 7–8 months back and was painless (Figs. 1, 2). On review of literature, only 1 more case with hemangiopericytoma in anterior maxilla of such young patient has been documented [12].

The aetiology of HPC remains unknown but several factors including trauma, long-term steroid use and arterial hypertension have been suggested as predisposing factors to this pathology [13, 14]. The patient also had history of trauma 1 year back which might have served as predisposing factor to the same.

On histopathology, it is usually fairly well circumscribed and exhibits tightly packed cells that surround endothelium-lined vascular channels. HPCs resemble too many spindle

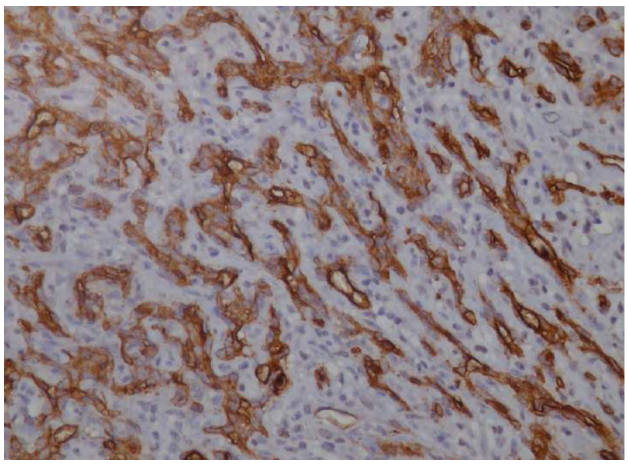

Fig. 4 Immunohistochemical staining

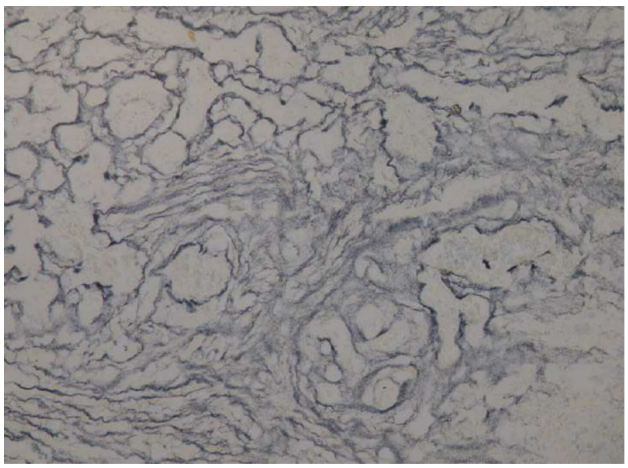

Fig. 5 Reticulin staining positivity in the vascular sheaths of the tumour (×40 view)

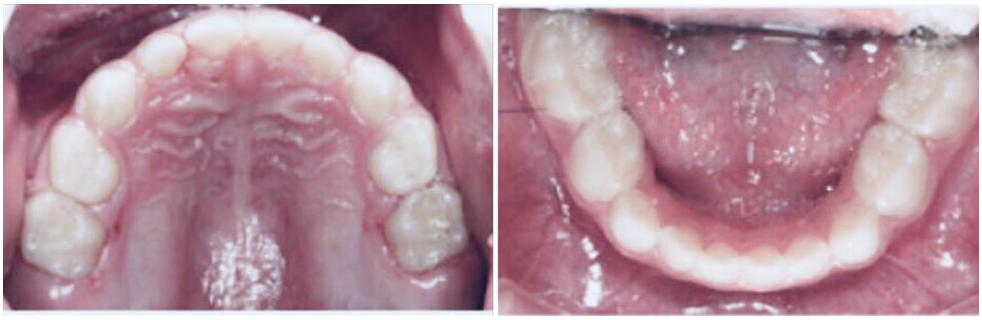

Fig. 6 Post-operative follow-up of 1 year

cell tumours. The so-called stag horn sign formed by proliferation of fusiform to roundish undifferentiated tumour cells in dendritic branches around the capillary vessels is considered prime diagnosis of HPCs [15]. Similar histopathological findings helped us to diagnose this 4-year-old child case with hemangiopericytoma (Fig. 3).

Also, hemangiopericytoma has a particular immunohistochemical profile, which aids in its diagnosis. In immunohistochemical findings, reticulin stain demonstrates a dense reticulin network that surrounds the vessels and individual tumours [16]. Actin and vimentin stains are also considered to be diagnostic findings which were present in our case (Figs. 4, 5).

The potential for malignant behaviour based on histological evaluation is often difficult to predict. Malignant lesions tend to demonstrate a lack of circumscription and high mitotic rate, and manifest a tendency towards necrosis, haemorrhage and invasion. In our case, mitotic figures were less than one per ten high power fields. There was no evidence of necrosis or local invasion and low-grade pleomorphism was the only dysplastic change that was observed. Even in the follow-up phase of 1 year, no metastatic changes were observed (Fig. 6).

The prognosis of HPC is unpredictable and its benign or malignant nature is unpredictable, and its benign or malignant nature is not easily discernible histologically in the first time [17]. The treatment of choice is wide local excision, and follow-up for long term is needed [18]. In our case, excisional biopsy was performed and until next 1 year, no recurrence was observed.

So with this case report we can conclude, although the initial impression can be that of any innocuous lesion such as gingival hyperplasia, a thorough clinical and histopathological examination is much needed to further avoid any aggressive state of solitary fibrous tumours or vascular tumours like HPC; latest diagnostic procedure such as immunohistochemical analysis ought to be performed.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical Statement The work described in our article reporting a case of hemangiopericytoma in a 4-year-old child has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki), and the appropriate consent was taken from the patient's guardians for the excisional biopsy performed. Also, the work conducted was approved by the Ethical Committee of the Institutional Board.

Informed Consent Informed Consent was taken from the patient's guardian for being the part of research work.

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