CASE REPORT

Hemangiopericytoma – An Unusual Case of the Maxilla and Mandible

Nandita Mohan[1], Akash Tripathi[2], Jayesh Anand[3], Saumya Singh[1], Rishvee Prateek[4], Md. Sibgatullah[5]

ABSTRACT

Hemangiopericytoma is a rare soft-tissue disease basically originating from the pericytes present in the capillary walls. Highlighting a case of a 4-year-old female patient presented with enormous gingival overgrowths in relation with maxillary palate and mandibular buccal gingiva. The diagnosis was confirmed with medical history and clinical findings. Surgical intervention was carried out as there was no change noted from medical management.

Keywords: Fibrous tumor, Hemangiopericytoma, Soft-tissue sarcoma

How to cite this article: Mohan N, Tripathi A, Anand J, Singh S, Prateek R, Sibgatullah M. Hemangiopericytoma – An Unusual Case of the Maxilla and Mandible. Int J Prev Clin Dent Res 2018;5(1):S199-202.

Source of support: Nil

Conflicts of interest: None

INTRODUCTION

A hemangiopericytoma (HPC) is a very rare, slow growing tumor of the soft-tissue that originates from the pericytes present in the walls of the blood capillaries, constituting <1% of all neoplasm. It is an uncommon tumor of infants. Pericytes, also known as the Zimmermann's pericytes, are contractile cells with long processes which wrap itself around the capillaries and serves to change the caliber of their lumens. Therefore, HPC have a wide distribution in both soft-tissue and skeletal system.

Although generally considered to be benign, distant metastases can occur.[1] The first description of this tumor was provided by Stout and Murray[2] in 1942. On first diagnosis, it is generally difficult to distinguish between benign and malignant forms. This frequently becomes evident only as the disease progresses. It is mostly observed that histopathological evaluation confirms the diagnosis. On histopathological examination, the tumor is characterized by proliferation of oval and spindle-shaped pericytic cells. Its incidence is mostly reported in the trunk or lower extremities.[3] However, the occurrence in the head and neck region represents 16–25% of all the reported HPC and the tumor represents 2–3% of all soft-tissue sarcomas in humans.[4] In this site, it affects soft tissues around the oral cavity, sinus tract, and meninges.[5]

Here is a discussion of one such case wherein the tumor involved both the maxilla and the mandible.

CASE REPORT

A 4-year-old girl reported to the Department of Paedodontics and Preventive Dentistry in the month of December, 2014, with the chief complaint of swelling in the gums of both the upper and the lower jaws since 8 months [Figure 1].

History of presenting illness revealed that the lesion initially presented as pea sized which rapidly increased to reach the present size. History of trauma noted 1 year back when the patient fell from the terrace, fell unconscious with bleeding experienced from the lateral part of the head and oral cavity for which she underwent a computed tomography scan where no positive radiographic findings were observed following which she was managed conservatively through medication for 2 months from a Government Hospital.

On further investigation, the parents gave a history of epilepsy since the age of 2 years, for which she was on medication as prescribed from a local Hakim. The frequency of the seizure was for about 5 min every 10 days. The bouts occurred at any time of the day. Post trauma, the patient is still under medication (Phenytoin) advised from a general practitioner. Since then, no seizure attacks were experienced.

[1,5]Consultant, [2,3]Senior Lecturer, [4]Post Graduate Student

[1]Department of Paedodontics and Preventive Dentistry, Sudha Rustagi College of Dental Sciences and Research, Faridabad, Haryana, India

[2]Department of Periodontology, Hazaribag College of Dental Sciences and Hospital, Demotand, Hazaribag, Jharkhand, India

[3]Department of Oral and Maxillofacial Surgery, Hazaribag College of Dental Sciences and Hospital, Demotand, Hazaribag, Jharkhand, India

[4]Department of Prosthodontics and Crown & Bridge, Dental College Azamgarh, Azamgarh, Uttar Pradesh, India

[5]Department of Oral and Maxillofacial Surgery, The Oxford Dental College and Hospital, Bengaluru, Karnataka, India

Corresponding Author: Dr. Nandita Mohan, Consultant Paedoodntist, 49, First Floor, Chitra Vihar, New Delhi - 110 092, India. e-mail: mohan.nandita@gmail.com

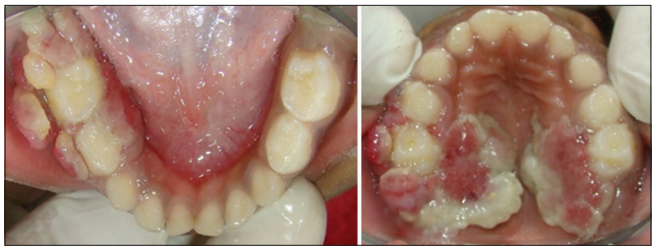

Figure 1: Pre-operative clinical photographs of both arches

Clinical Examination

General physical examination showed normal gait and moderate built. Extraorally, there was no facial asymmetry; however, left submandibular lymph nodes were tender and palpable. Intra oral findings revealed a primary dentition stage with the presence of proliferative, lobulated ulcerated pedunculated growths of the gingiva seen wrt 74, 75 region on the buccal aspect and on palatal aspects of the maxilla in the region of 54, 55 and 64, 65, extending from the marginal gingivae of the mentioned teeth, having a firm base, to the center of the palate covering the entire palatal vault. It had progressed from a pea size to approximately 3 cm×4 cm without crossing the midline.

On clinical examination, the overlying mucosa appeared erythematous covered with slough. However, on palpation, it appeared soft, fluctuant and rubbery in consistency, non-tender, and non-pulsatile.

Provisional Diagnosis

Keeping the history of epilepsy, a provisional diagnosis of drug induced gingival enlargement was made. However, malignant neoplasms of the hard palate including ossifying fibroma, fibrosarcoma, Burkitt's lymphoma, Ewing's sarcoma, chondrosarcoma, and hemangioendothelioma, and HPC were the differential diagnosis. Relevant radiological and histopathological investigations were undertaken to confirm the aforementioned.

Radiographic Investigations

An orthopantomogram as well as each quadrant intraoral radiographs were advised to assess the changes in the normal pattern. However, no abnormalities were detected in the bone morphology and eruption pattern of teeth [Figure 2].

Other Investigations

Routine blood investigations along with ultrasound of the abdomen were done. Hematological report revealed normalcy but with a marginal decrease in red blood count, PCV, mean corpuscular volume, and mean corpuscular hemoglobin (4.31, 33.6, 78.0, and 26.2, respectively). The ultrasound showed no abnormality in the internal organs.

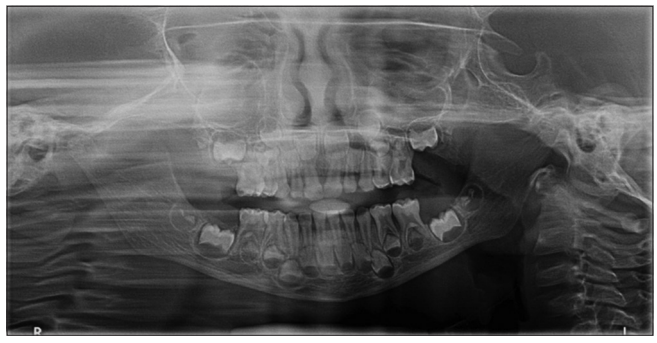

Figure 2: Ortho-pentomogram of the patient

Biopsy

Excisional biopsy of the whole lesion involving the maxilla and the mandible was performed under general anesthesia using electro cautery [Figure 3].

The bleeding was controlled and the tissue specimen was sent for histological analysis.

The histopathological report showed a highly cellular connective tissue stroma with numerous atypical spindle cells arranged in a lobular pattern at some places and in other areas present irregularly. Tumor cells exhibited the presence of various dysplastic features such as a high degree of cellular and nuclear pleomorphism, hyperchromatic, and even vesiculated nuclei with few mitotic figures. The tissue was highly vascular with numerous engorged blood vessels of varying size and shape. The blood vessels exhibited a STAG HORN pattern arranged in a form of slit-like spaces with the presence of small round indistinct lumen. Spindle shaped and plump tumor cells seemed to be lining the blood vessels and filling the stroma. There were scanty amount of inflammatory infiltrates chiefly consisting of lymphocytes and plasma cells, dispersed throughout the lesion. Connective tissue septae with dense collagenous fibers were observed. Overlying epithelium appeared atrophic to hyperplastic parakeratinized stratified squamous epithelium with elongated rete ridges at places. Superficial areas showed large amount of fibrinous exudates containing scattered chronic inflammatory cells. The histopathological features of the case were consistent with malignant HPC.

Final Diagnosis

On the basis of history and clinical, radiological and histopathological examinations a final diagnosis of HPC was made.

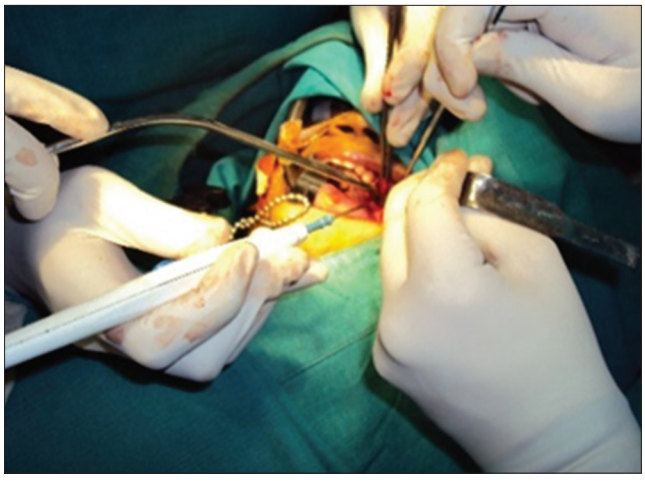

Figure 3: Excision of the lesions under general anesthesia

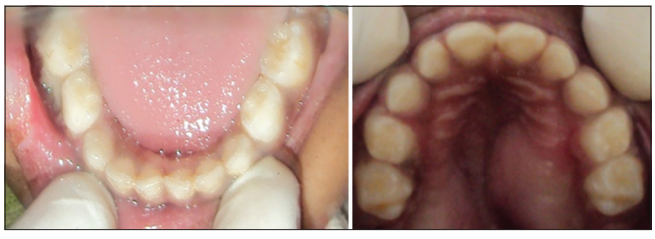

Figure 4: Post-operative clinical photographs of both arches

Follow-up

During the 1st month follow-up, the child remained asymptomatic [Figure 4].

However, in the next visit, after 3 months the lesion recurred as a pinpoint growth in the palatal aspect wrt 54 and 55. The patient is still under the follow-up protocol and no further change in the size of the growth is noticed since.

DISCUSSION

HPC mostly arises in the 4th–6th decade of life and is rarely seen in children. Pediatric cases account for <10% of all HPC.[6] However, it can either be benign or malignant.

Two subtypes have been described: (1) Infantile HPC occurring in infants <1-year-old; and (2) adult HPC in anyone older.[7] The etiology still remains unknown, although its presence is linked to trauma, prolonged steroid use, and/or hormonal imbalance, which were negative findings in our case because of which the diagnosis becomes a challenging task due to its rare occurrence and no specific clinical behavior.

Gingival overgrowth is a common side effect of phenytoin, termed "Phenytoin-induced gingival overgrowth."[8] Other drugs which cause gingival overgrowths include anticonvulsants such as phenytoin, phenobarbital, lamotrigine, vigabatrin, ethosuximide, topiramate and primidone, and calcium channel blockers, such as nifedipine, amlodipine, and verapamil and cyclosporine, an immune-suppressant.

Of all cases of drug-induced gingival overgrowth, about 50% are attributed to phenytoin as noted in our case, 30% to cyclosporins and the remaining 10–20% to calcium channel blockers.

HPCs may occur anywhere in the body, with the most common anatomic locations being the lower extremities, the pelvis, and the head and neck region. HPCs are usually deep seated and found in the muscle tissue; dermal and subcutaneous HPCs are much less common. It is a neoplasm which is usually benign but has a definite malignant counterpart. Under light microscopy, HPC is characterized by vascular channels arranged in staghorn manner.[9] 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. Histopathologically, all the features were noted in our case suggestive of a malignant condition.

Very few reports about its clinical management have been published till date. Therefore, the management of such lesions can vary but the surgical resection remains the treatment of choice, which was followed for the present case. However, other treatment modalities such as chemotherapy, giving high response rates have also been reported. Positive responses to chemotherapy have been reported with vincristine, cyclophosphamide, deoxorubicin, dactinomycin, methotrexate, mitoxantrone, and other alkylating agents as well. On the contrary, radiotherapy is rendered effective in some patients, but indications are limited because of the age factor.

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 at least 10 years.

CONCLUSION

The early diagnosis and management of these lesions can limit post-surgical morbidity. Careful follow-up is therefore essential. Child patient prognosis and long-term survival can vary greatly from individual to individual. Therefore, prompt medical attention and aggressive therapy are the important aspects to achieve best prognosis for such individuals.

REFERENCES

1. Bailey PV, Weber TR, Tracy TF, O'Connor DM, Sotelo-Avila C. Congenital hemangiopericytoma: An unusual vascular neoplasm of infancy. Surgery 1993;114:936-41.
2. Stout AP, Murray MR. Hemangiopericytoma: A vascular tumor featuring Zimmerman's pericytes. Ann Surg

1942;116:26-33.
3. Espat JN, Lewis JJ, Leung D, Woodruff JM, Antonescu CR, Shia J, *et al*. Conventional hemangiopericytoma. Modern analysis of outcome. Cancer 2002;95:1746-51.
4. Rajendran R, Sivapathasundharam B. Benign and malignant tumors of the oral cavity. In: Shafer`s Oral Pathology. Netherlands: Elsevier; 2009. p. 162-3.
5. Carvalo JR, Haddad L, Leonhardt FD, Filho MF, de Oliveira Santos R, Cervantes O, *et al*. Head and neck hemangiopericytoma in a child: Case report. Sao Paulo Med J 2004;122:223-6.
6. Brar R, Kulkarni S, Sheikh S, Jindal S, Brar P. Hemangipericytoma associated with multiple keratocystic odontogenic tumors in an adolescent patient: A case report. J Oral Sci 2008;50:233-7.
7. Anand R, Gupta S. Hemangiopericytoma of the maxilla in a pediatric patient: A case report. J Dent Child 2010;77:180-2.
8. Arya R, Gulati S. Phenytoin-induced gingival overgrowth. Acta Neurol Scand 2012;125:149-55.
9. Archana J, Gupta SK. Sinonasal hemangiopericytoma. Bombay Hosp J 2008;50:660-63.