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Hemangiopericytoma– Rarest of the rare at uncommon site

Sumit Asrani[1], RB Dhirawani[2], Sanchit Jain[3], Indraj Arora[4,*]

[1,3]Senior Lecturer, [2]Professor & HOD, [4]Resident, Hitkarini Dental College, Jabalpur, Madhya Pradesh

***Corresponding Author:**
Email: indrajarora09@gmail.com

Abstract

Hemangiopericytoma are vascular lesions are more commonly reported in the soft tissues when compared to reports in the hard tissues. These are considered to originate from the pericytes in the blood capillaries and hence can occur anywhere in the human body. The incidence of this entity among the other vascular lesion is only 0.1%. The most common locations reported are the brain, lower extremities, pelvic area, head, and neck. Hemangiopericytoma is most commonly located in the nasal cavity and paranasal sinuses in the head and neck region. Hemangiopericytoma are painless masses and may not have any associated symptoms but may turn symptomatic when other adjacent structures are affected. Though rare, their prognosis is better. This is because of the tendency to be less aggressive and not metastasizing. Here we attempt to report this pathology at zygomatic bone which is the rarest site of the occurrence of the vascular lesions as seen in literature.

Keywords: Hemangiopericytoma, Weber ferguson, Pericyte

Introduction

Hemangiopericytoma is a relatively rare neoplasm of vascular origin that may manifest itself in head and neck region. Hemangiopericytoma can occur anywhere in the body where there is presence of capillaries and hence can be present in any part of the body. This pathology as an entity is described under perivascular tumours which most commonly affects the nasal cavity and paranasal sinus in head and neck region. The incidence was first confirmed by a study conducted by Enzinger and Smith in 1976 where they evaluated a total of 106 patients. The study concluded that the incidence in head and neck was only 16 % which is further confirmed with recent studies.[1,2] Hemangiopericytoma is relatively rare entity & thought to arise from Zimmerman cell, a population of contractile cells which regulate the calibre of capillaries whose discussion and detailed description has been found in literature ever since the first description of this entity by Stout and Murray in the year 1942.[3,4] Various other authors also performed detailed study on this pathologic entity and commented that it occurs in lower extremities or pelvis in adulthood. The incidence of hemangiopericytoma is only 1% amongst the vascular tumours which further determinates the rarity of this entity.[2,3] This pathological disease is mainly diagnosed on the basis of the histopathological grounds.[4,5] The clinical presentation of this disease is highly variable making clinical diagnosis of this disease near impossible in conjugation to variable clinical differential diagnosis which could be made in accordance to presentation. There has been reports of hemangiopericytoma involving the mandible, palate, buccal mucosa and lip.[4,6] We are presenting here a case of hemangiopericytoma along with detail description of its features which was done at our centre.

Case Report

A 13 year old female patient was referred to our department of oral and maxillofacial surgery from a peripheral centre for diagnosis and treatment. The patient had a chief complaint of swelling on the right side of the face which was painful and was progressively growing in size. On detailed history taking patient's parents informed that the swelling was present on her face since the age of 8 years. The patient had earlier visited other centres and had undergone treatment for the same swelling twice. Patient was unable to provide with exact details of the procedure which was carried out during the previous interventions due to lack of record keeping. The swelling was constantly growing at a slow pace and gradually increased to the extent as shown in Fig. 1. There was pain associated with the swelling which was dull and continuous. The pain was non-radiating in nature which was relieved on medications. There were no associated aggravating or relieving factors which were found in association with the pain.

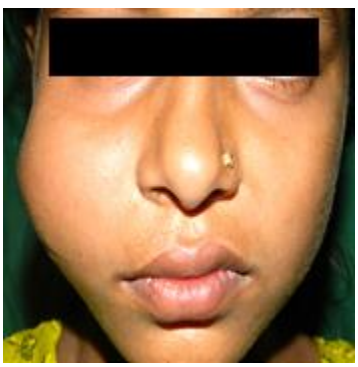


Fig. 1: Frontal View

Clinical examination of the swelling revealed the presence of a single solitary swelling present on the

right side of the face which was dome shaped. This swelling extended super-inferiorly from the infraorbital region till the corner of mouth and anterio-posteriorly on the malar prominence of the zygomatic bone. The skin overlying the swelling had scar of previous surgical intervention which was evident clinically. All the findings seen on inspection were confirmed on the palpation. The margins of swelling were well defined and the swelling was non tender on palpation. The swelling was firm in consistency and non-mobile. There was no associated active discharge. Intraoral examination revealed obliteration of buccal vestibule from 13 to 16 region which was due to extension of the swelling. All the findings on palpation were similar to palpation of extraoral swelling.

We attempted for fine needle aspiration cytology (FNAC) to evaluate the swelling but there was no aspirate hence FNAC was inconclusive. This was followed by performing incisional biopsy, the reports of which were conclusive of hemangiopericytoma.(Fig. 2) The vascular malformations at zygomatic region is relatively rare as found in literature earlier making it even more rare presentation.[7] The histopathological report hence was confirmed again at higher centre for the same.

The patient was advised computed tomography with contrast enhancement and 3-D reconstruction. The CT scan revealed the exact extent of the pathology along with the location of lesion at an uncommon site being zygoma. The lesion observed was having a irregular surface with distinct well defined margins which demarked the lesion from normal bone. The cortical bone was eroded as observed and axial cut of the CT scan revealed involvement of medullary bone of the malar prominence of the zygomatic bone. The anterior extension of the lesion was along the zygomaticomaxillary suture and the posterior extent was about 2 mm ahead of the zygomaticomaxillary buttress and inferiorly involving the lower margin of the zygomatic bone(Fig. 3). This clinical entity reported to have highrecurrence rate and hence was treatment aggressively. The treatment plan involved wide surgical excision of the pathology with clear resection margins.

The relevant features which determined the swelling to be hemangiopericytoma seen histo-pathologically are also shown in the Fig. 2. Slit-like spaces surrounded by pericytes, capillary-like tubules, lined by a single layer of flattened endothelial cell and the characteristic "stag-horn" appearance of hemangiopericytoma are well demonstrated histopathologically.[6,8]

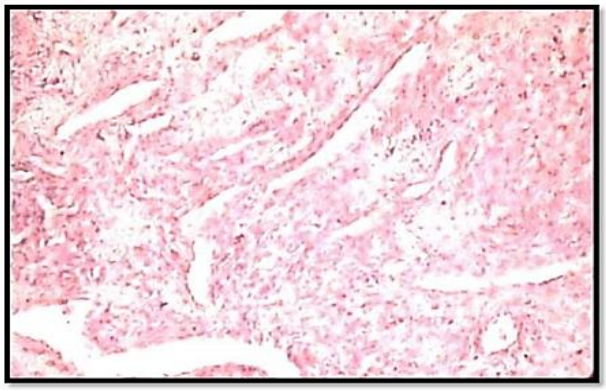


Fig. 2: Histo pathology slide

The modified Weber Ferguson incision including the infraorbital incision was used for adequate exposure of the maxilla and zygoma(Fig. 4 & 5). The affected bone was excised along with a wide and adequate margin of healthy bone. The excision included removal of inferior border of the zygomatic bonealong with the removal of part of zygomatic malar prominence in both vertical and horizontal dimensions (Fig. 6). The surface morphology of the growth which was evident on exposure was rough. It presented itself as a protruding reddish-gray mass with marked bleeding on contact which was similar to description giver earlier in literature.[2,6] The maxillary bone and adjoining structures were not involved and here were preserved adequately which does not affect the facial aesthetics post-operatively.

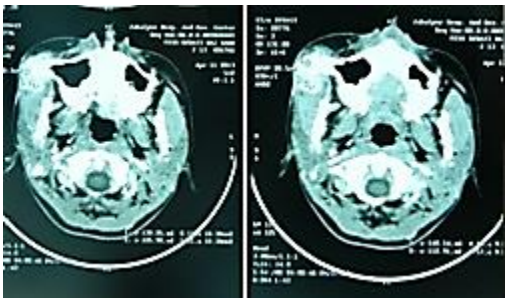


Fig. 3: CT Axial cut

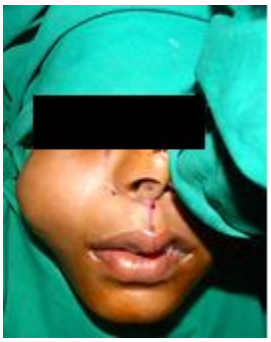


Fig. 4: Incision Marking

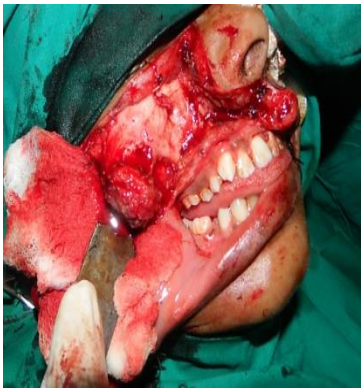


Fig. 5: Exposure

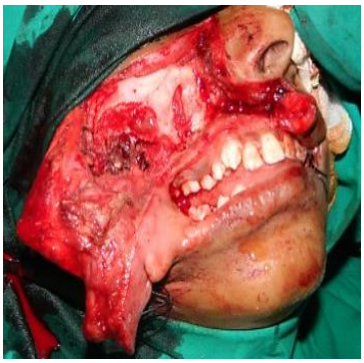
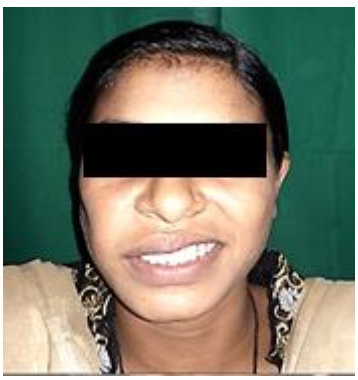


Fig. 6: Excision of lesion

The patient has been on regular follow-up for the past 2.5 years. There have been no signs of recurrence in terms of any post-surgical swelling, pain or any other discomfort associated with the region where the excision of the lesion was performed. There was no intraoral defect. The extra-oral incision also healed adequately and with aesthetically acceptable scars. Patient has not complain regarding her aesthetics hence no secondary surgery performed for the reconstruction of the defect.(Fig.7)



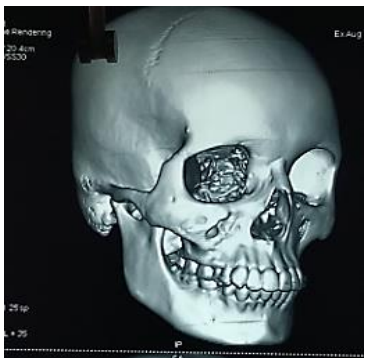


Fig. 7: Post operative frontal picture and CT cut (3D Reconstruction)

Discussion

The literature regarding the hemangiopericytoma are relatively scant and mainly includes case reports or case series. Hemangiopericytoma are believed to have incidence which is independent of sexes and all age groups.[1,2] In 2002, WHO classification of tumours this entity was regarded as a tumour with potential low malignancy rather than classifying it under benign or malignant tumour. The clear diagnostic criteria for grading of malignancy of these lesions were not included in WHO classification.[9]

Histological criteria for tumour malignancy are less predictive with respect to the behaviour of hemangiopericytoma. The histopathological grading in itself determines the aggressiveness of any pathology which has to be considered for the treatment planning. They can be divided in to three grades being low (Grade I), intermediate (Grade II), high (Grade III). The various features which are considered for grading includes the mitotic index, cellularity, cytoplasm, nuclei, chromatin.[10] Though histopathological grading was not done in our case, owing to the recurrence aggressive surgical resection was carried out in our case.

There is relative rare incidence of intraosseous vascular malformations involving the zygoma bone. The case presented in this case report is an intraosseous hemangiopericytoma which has affected the malar portion of the zygomatic bone.[7] This relatively less incidences of vascular lesions in zygomatic region makes the reported case unique.

Hemangiopericytoma involving the bones are divided into primary which may be intramedullary and periosteal or secondary forms. Tumours which initiates in the medullary cavity causes expansion and destruction of the cortex which later extends into the soft tissues.[7,9] The presented case is of intramedullary type which is evident from the CT scan images.

Radiation therapy is not used, primarily because of the radio-resistant nature of the tumour.[2] The tumour is highly vascular and hence adequate management is necessary in terms of surgical excision of the same. The wide surgical excision of the pathology is

recommended treatment owing to its potentially aggressive behaviour and long term follow-up is advised to ensure the adequacy of treatment done.[6] The reported recurrence rate of this pathology ranges from 7% to 20%, with an average time of recurrence of 6–7 years. There are even reports of recurrences of this pathology with rates more that 50%.[6,11]

The reconstruction of the defect can be carried out after 6 months in highly aggressive tumours where as in low grade it can be carried out in immediate phase as the recurrences are high only after 6-7 years which is a long duration.[5,6,11]

Conclusion

Hemangiopericytoma is a relatively rare tumor in occurrence in head and neck region with reporting of such lesion extending intramedullary in the zygomatic bone once only in literature. The case presented here is hence unique and signifies the importance of proper histopathological evaluation of any pathology which planning for treatment. It is mandatory for complete clinical evaluation of any case before hand.

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