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Hemangiopericytoma/solitaryfibrous tumor of mandible: A rare entity

Samapika Routray, Neeta Mohanty, Swagatika Panda, Sujit Ranjan Sahoo
Department of Oral Pathology and Microbiology, Institute of Dental Sciences, Siksha'O' Anusandhan University, Ghatikia, Bhubaneswar, Orissa, India

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INTRODUCTION

The hemangiopericytoma (HPC) is a tumor of the pericytes. It represents 1% of vascular tumors.[1] The fourth edition of the World Health Organization (WHO) classification of tumors of soft tissue and bone "blue book" that was published in February 2013 has abandoned the term "HPC." It is used only to describe a morphological pattern that is shared by different entities. Currently, solitary fibrous tumor (SFT), HPC, lipomatous HPC and giant cell angiofibroma are all grouped under the "extra-pleural SFT" category.

CASE REPORT

A 54-year-old female patient reported to The Dental Hospital with a history of a small swelling in her left ramus since 2 months. Extra-orally a single, diffuse swelling was seen in the left ramus of the mandible. A well circumscribed, lobular growth measuring about 4 cm × 2 cm in size in the left alveolar region was seen during intra-oral inspection. The swelling was extending from the retromolar region to the second premolar region obliterating the buccal sulcus, with bicortical expansion posteriorly. It was firm in consistency, expanding and extending below the inferior border of the mandible and is nontender on palpation. Teeth involved in the swelling were mobile. Fine-needle aspiration cytology was performed, revealing a scanty serosanguinous fluid. Panoramic radiograph revealed a single, multilocular radiolucency in the left ramus extending anteriorly upto the first molar, the corticated margin of the lesion extended beyond the inferior border of the mandible along with root resorption of the first molar.

The hematoxylin and eosin stained section showed:

- Endothelial lined blood vessels with perivascular hyalinization in the background of a highly cellular and uniform appearing delicate, collagenous connective tissue stroma [Figure 1,×100]
- On higher magnification, the cells were mostly plump, short, spindle in appearance with mild cellular atypia and were arranged in typical whorling pattern [Figure 2, ×400]
- Areas showed typical staghorn appearance of blood vessels [Figure 3, ×100] and on higher magnification these blood vessels showed plump endothelial cell lining [Figure 4, ×400]
- Heterotopic ossification was observed in the background of a delicate collagenous stroma, which can be related to the pluripotency of the adjacent plump spindle cells [Figure 5, ×200].

The positive silver reticulin stain [Figure 6] and CD34 immunohistochemical marker [Figure 7] concluded the diagnosis of HPC.

Differential diagnosis

- Mesenchymal chondrosarcoma has to be differentially diagnosed as an HPC with cartilaginous differentiation is also being described. Previously this lesion was described as mesenchymal chondrosarcoma[2,3]
- The so-called cleft formation and biphasic appearance of the synovial sarcoma should be carefully looked for to differentiate from prominent vascular channels in hemangiopericytoma.[2,3] CD34 negativity was a reason for excluding monophasic synovial sarcoma[4]
- Reticulin stains readily differentiate the proliferation of endothelial cells, as seen in the cellular

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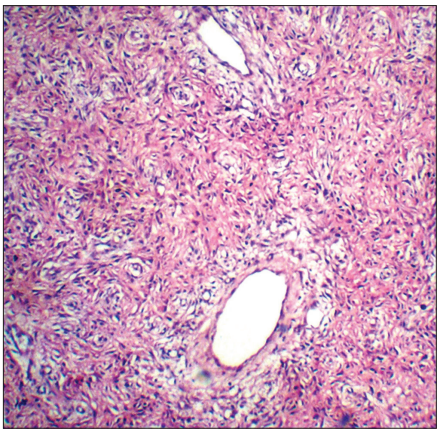

Figure 1: Endothelial lined blood vessels with perivascular hyalinization in the background of a highly cellular and uniform appearing delicate, collagenous connective tissue stroma (H&E stain, x100)

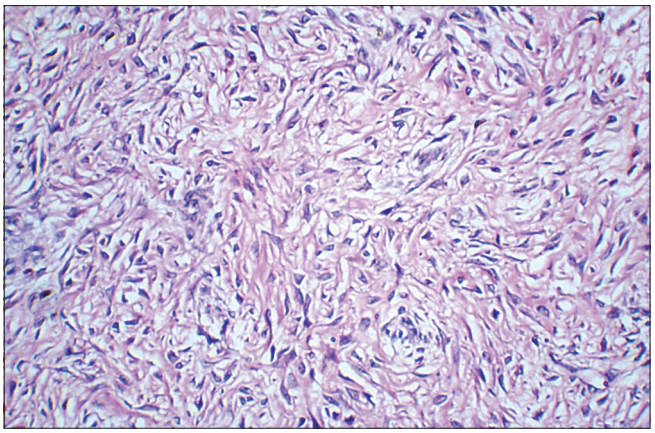

Figure 2: Higher magnification showing arrangement of cells in typical whorling pattern (H&E stain, x400)

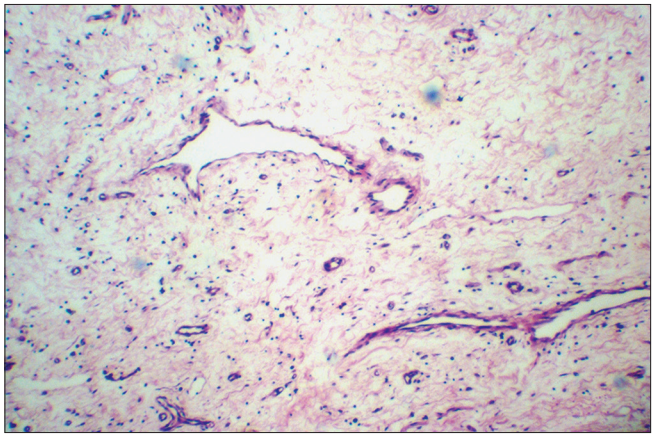

Figure 3: Typical staghorn appearance of blood vessels (H&E stain, x100)

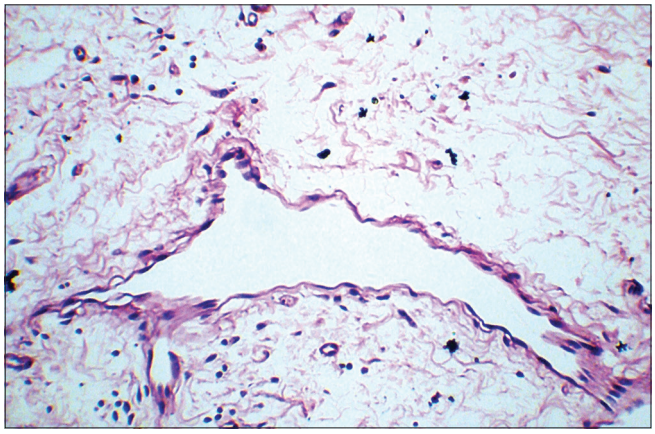

Figure 4: Higher magnification these blood vessels showed plump endothelial cell lining (H&E stain, x400)

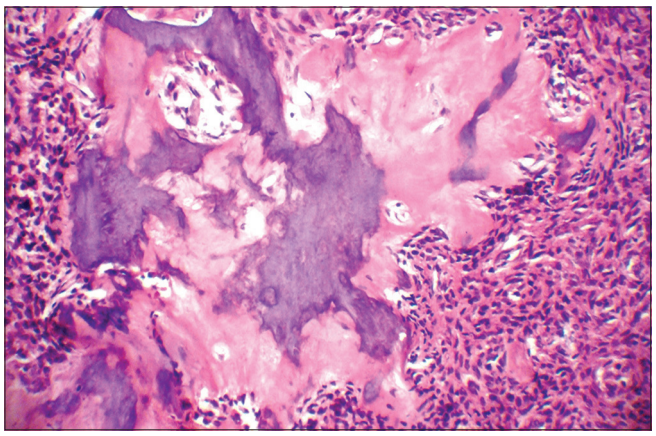

Figure 5: Heterotopic ossification in the background of plump proliferating fibroblasts. (H&E stain, x200)

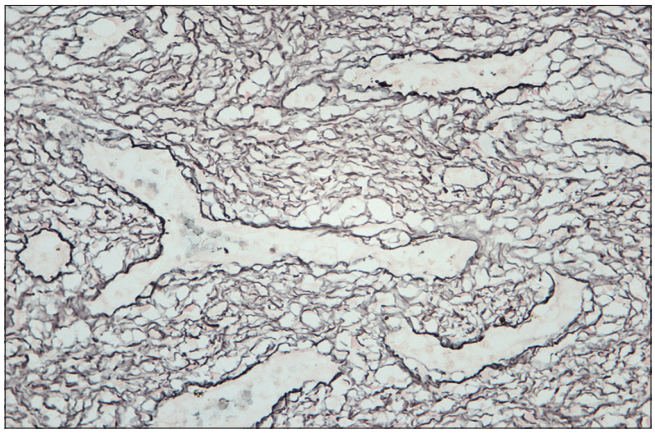

Figure 6: Silver reticulin stain showing reticular fibers surrounding the tumor cells outside the vessel wall. (Silver stain, x100)

hemangioendothelioma. The endothelial cell does not produce reticulin and presents as cellular clusters, devoid of reticulin within the vascular spaces, in contrast to the extra-vascular distribution of reticulin separated pericytes of the hemangiopericytoma

- Glomus tumor and Kaposi's sarcoma are also considered in the differential diagnosis of hemangiopericytoma[2,3]
- It is exceedingly difficult to differentiate benign fibrous histiocytoma from hemangiopericytoma.[2,3] Malignant fibrous histiocytoma should be excluded after careful observation of nuclear activity, mitotic activity and tumor necrosis.[5] Both benign and malignant fibrous histiocytoma should primarily be excluded when there is absence of histiocytic like cells.

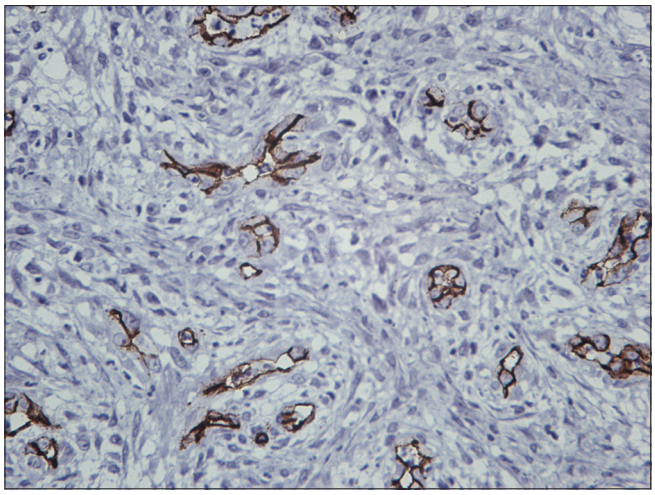

Figure 7: The endothelial cells showing positivity for CD34 (IHC stain, x100)

DISCUSSION

After the description of SFT by Klemperer and Rabin, many pathologists opine that SFT and HPC are two spectrums of the same disease based on histologic similarities between the two.[6] However, in 2013 by WHO standards, they were included under the heading of extra-pleural SFT. The cellular end of this spectrum corresponds to classic HPC whereas the hyalinized end is referred to classic SFT. The features of classic HPC consists of tightly packed round to fusiform cells arranged around an elaborate vasculature. Typically, the sinusoidal vessels have a staghorn pattern. Myxoid change is also common. In contrast, lesions having features of classic SFT show "patternless pattern." Another characteristic feature is intense hyalinization resulting in gaping staghorn vessels. The lineage of these two lesions and terminologies still remain debatable. Taking into consideration, the comparison of HPC and SFT as mentioned by Weiss *et al.*[3] we inferred our diagnosis. SFT has a mesenchymal phenotype and characteristically overexpress CD34, a myeloid precursor cell antigen seen in normal and neoplastic endothelial cells[7] and in some primitive mesenchymal cells. CD34 expression in neoplastic cells is the most reliable and consistently positive marker of SFTs at both pleural and extra-pleural sites.[8-10] Furthermore, supporting the findings of Westra *et al.*,[11] we can conclude that CD34 immunoreactivity is a sensitive marker for SFT and in conjunction with an appropriate immunohistochemical panel CD34 is useful in discriminating SFTs from other histologically similar neoplasms. The parameters used for comparison are described in Table 1. Thus, these features helped us to diagnose the lesion as HPC/SFT of the mandible, making it a rarity due to the site presentation.

Final Diagnosis: SFT/Hemangiopericytoma.

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Nil.

Table 1: Conclusive features pertaining to diagnosis of the case

Parameters	HPC	SFT	Case
Location	Usually extremity	Usually body cavity, particularly pleura	Mandible
Pericytic vascular pattern	+++ (definitional)	focal	Reticulin strongly positive
Broad zone of hyalinization	Variable to focal	Typical	restricted to perivascular areas only
CD 34	Focal positivity	Diffuse	Focal
MIB 2	Intensely positive	Less strong	Intensely

HPC: Hemangiopericytoma, SFT: Solitary fibrous tumor

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Güerrissi JO, Miranda MG, Olivier R. Giant hemangiopericytoma of mandible: A propos of a case: A variant of the surgical technique for protection of the articular fosse. J CraniofacSurg 2006;17:523-7.
2. Gnepp DR. Diagnostic Surgical Pathology of the Head and Neck. 2nded. Philadelphia: Elsevier; 2009.
3. Weiss SW, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors. 5thed. Philadelphia: Mosby Elsevier; 2008.
4. Alsharief AN, Fageeh M, Alabdulkarim Y. Monophasic synovial sarcoma presenting as a primary ileal mass: A case report and review of the literature. J Med Case Rep 2012;6:83.
5. Pilch BZ. Head and Neck Surgical Pathology. 1st edition. Maryland, USA: Lippicott Williams and Wilkins publications; 2000.
6. Klemperer P, Rabin CB. Primary neoplasm of the pleura: A report of five cases. Arch Pathol 1931;11:385-412.
7. Traweek ST, Kandalaft PL, Mehta P, Battifora H. The human hematopoietic progenitor cell antigen (CD34) in vascular neoplasia. Am J ClinPathol 1991;96:25-31.
8. Bost F, Barnoud R, Peoc'h M, Le Marc'hadour F, Pasquier D, Pasquier B. CD34 positivity in solitary fibrous tumor of the liver. Am J SurgPathol 1995;19:1334-5.
9. Flint A, Weiss SW. CD-34 and keratin expression distinguishes solitary fibrous tumor (fibrous mesothelioma) of pleura from desmoplastic mesothelioma. Hum Pathol 1995;26:428-31.
10. Miettinen M, Lindenmayer AE, Chaubal A. Endothelial cell markers CD31, CD34 and BNH9 antibody to H- and Y-antigens – Evaluation of their specificity and sensitivity in the diagnosis of vascular tumors and comparison with von Willebrand factor. Mod Pathol 1994;7:82-90.
11. Westra WH, Gerald WL, Rosai J. Solitary fibrous tumor. Consistent CD34 immunoreactivity and occurrence in the orbit. Am J SurgPathol 1994;18:992-8.