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Original Research Paper

Pathology

CD 34 NEGATIVE SOLITARY FIBROUS TUMOR PRESENTING AS MANDIBULAR SWELLING- A RARE CASE REPORT

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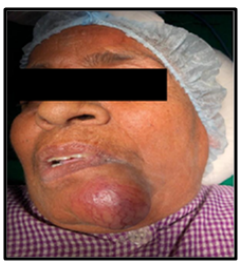
ABSTRACT Solitary fibrous tumors (SFTs) are uncommon mesenchymal neoplasms of fibroblastic type that can arise anywhere in the body. Recently, NGF1-A binding protein 2 (NAB2)- signal transducer and the activator of transcription 6 (STAT6) fusion gene were discovered as hallmark of SFTs/ HPCs by using whole- exome and transcription sequencing. Consequently, the fusion gene can be rapidly detected by STAT 6 IHC.

KEYWORDS : 34 NEGATIVE SFT, TLE-1, STAT-6

INTRODUCTION

SFT were firstly reported in the pleura by klempner and ratin in 1931. These tumors are rare lesions and although formerly believed to be restricted to the pleura, tumors showing features of SFT have been increasingly recognized in extrapulmonary sites. SFTs arising in the soft tissue of head and neck account for 10% of all cases. Those occurring in oral cavity accounts for 3% of all head andneck cases. In the present report,we describe the occurrence of SFT presenting as an extraskeletal mandibular swelling.

67 year old female came with complaints of gradually increasing painless non tender swelling over left mandible since 1 year**. On examination**, swelling was partially mobile with restricted mobility in all directions - The clinical diagnosis was suspicious of sebaceous cyst.



SWELLING OVER LEFT MANDIBLE

The **USG findings** revealed well defined heterogeneously hypoechoic lesions measuring 3.7x 3cm with hyperechoic areas noted at the site of swelling over the body of mandible.- Features were suggestive of neoplastic etiology

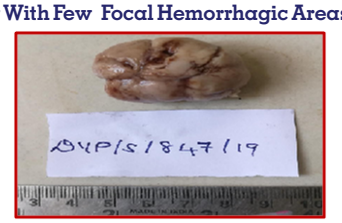
Fine needle aspiration cytology (FNAC) was performed showing features suggestive of spindle cell neoplasm.

Histopathology for confirmation was advised.

Surgery was performed, the swelling was in subcutaneous plane with no attachment to underlying structures.

Histopathology sample recieved showed well encapsulated and lobulated soft tissue mass of size measuring 4x3x1.5cm with congested areas. On serial sectioning – C/S- Solid, homogenous, white, soft to firm in consistency with few focal hemorrhagic areas.

Cut Section- Solid, Homogeneous White, Soft To Firm In Consistency With Few Focal Hemorrhagic Areas.



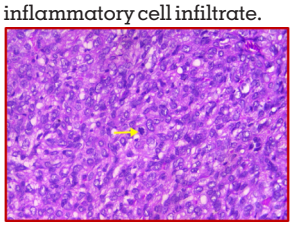
Well Encapsulated And Lobulated Soft Tissue Mass Of Size 4x3x1.5 Cm With Congested Areas.

Histological findings- Tumor mass showed monomorphic cells arranged in streaming fascicle with plenty of branching thin walled blood vessels lined by endothelial cells giving the appearance of hemangiopericytic vascular like pattern.

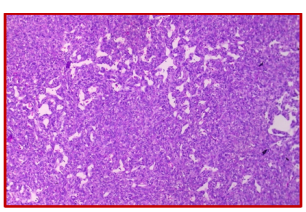
Individual tumor cells were large with epithelioid to spindle in shape with rounded ends, vesicular nuclei, coarse chromatin, inconspicuous to prominent nucleoli.

Cytoplasm was scant and clear to eosinophilic with interspersed pink collagen bundles with focal myxoid areas seen.(perivascular hyalinization).

Few mitotic figures seen. No necrosis, hemorrhage seen.No other specific pattern was seen. No tumor giant cells and inflammatory cell infiltrate.



H&e- 100x Magnification Yellow Arrow- Shows Abnormal Mitotic Figure



H&e- 40x Magnification Hemangio- Pericytomatous Pattern

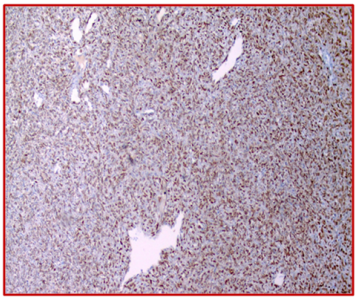
IHCFINDINGS-

Positive for

- EMA (Focal)
- BCL2
- CD 99(MIC2)
- INI1:Reduced expression
- **STAT-6**
- Ki 67

Negative for

- CK
- Desmin
- SMA
- **CD 34**
- S100
- P63
- **TLE-1**



STAT- 6- STRONG NUCLEAR POSITIVITY

DISCUSSION-

CD- 34 expression is found in most cases of SFTs therefore it is regarded as a useful positive marker.

However, CD- 34 expression can be lost in 5-10% of SFTs which is seen in our case.

CD 34 is not entirely specific since it can be expressed in a variety of other mesenchymal tumors, therefore a more sensitive markers like STAT 6 along with CD99 should be used. BCL- 2 could provide more supportive diagnostic information in differential diagnosis of SFT from other spindle cell neoplasms. Theabsence of S-100 is essential for ruling out myogenic, PSNT, fibroblastic and fibro-histiocytic neoplasm with spindle cell features.

The differential diagnosis may prove difficult since synovial sarcoma rarely express CD 34, and CD 99 can be immunohistochemically detected in 60-70% of synovial sarcoma. Also, the molecular study SYT-SSX1 OR SYT-SSX2 fusion transcript is must to rule out SS.

But the absence of TLE-1 which is an excellent discriminator of synovial sarcomas from its other close differential sarcomas and expression of STAT- 6 nuclear positivity favour the diagnosis of SFT in our case.

CONCLUSION:

Differential diagnosis of SFT is very extensive. Hemangio-pericytomatous pattern is present in a wide variety of tumors and the epithelioid growth pattern contributes to further difficulties.

The main challenge is to differentiate it from synovial sarcoma (SS) if SFT expresses EMA and its rare expression of Cd34.

CD 34 expression is found in most cases of SFTs with few exceptions. However, its expression can be lost in histologically high grade tumors or tumors from patient with repeated recurrences. It is associated with malignant transformation and its loss has been documented in the cases with unfavourable outcome.

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