University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2020, Vol. 6(1)

A RARE CASE OF SYNOVIAL SARCOMA OF TONSIL

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Abstract : Synovial sarcoma rarely occurs in the head and neck region(3-5) . Literature reveals that primary tonsillar Synovial Sarcoma has previously been documented only in three patients (1-6) .It has generally been regarded as a high grade sarcoma. Recent analysis of clinical, morphological, and molecular characteristics of Synovial Sarcoma, however, identified low and high risk group of patients, resulting in important implications for the treatment of patients diagnosed with Synovial Sarcoma. In all of these patients the tumor was histologically biphasic.

Keyword :SYNOVIAL SARCOMA , TONSIL, LATERAL PHARYNGEAL WALL, HNSS

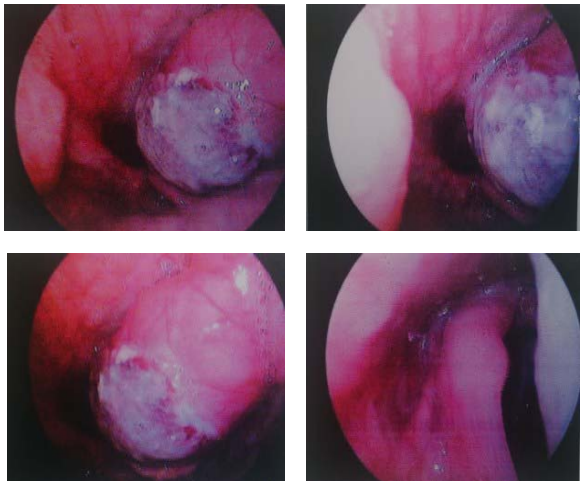
INTRODUCTION:

Synovial sarcoma is defined as mesenchymal spindle cell tumor that exhibits variable epithelial differentiation, including gland formation, and specific chromosomal translocation. It may occur at any site in the body, however only 3–5% of SS arise in the head and neck region (1,2) .It is thought to arise from pluripotent mesenchymal cells with both epitheloid and spindle differentiation. It has a bimodal age distribution with incidence peaking in the under fives and the adolescents. These tumours have a tendency to recur locally and metastasize. This paper is about the occurrence of this tumour in an adolescent girl.

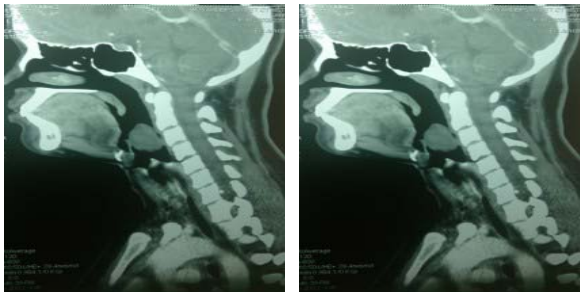
CASE HISTORY:

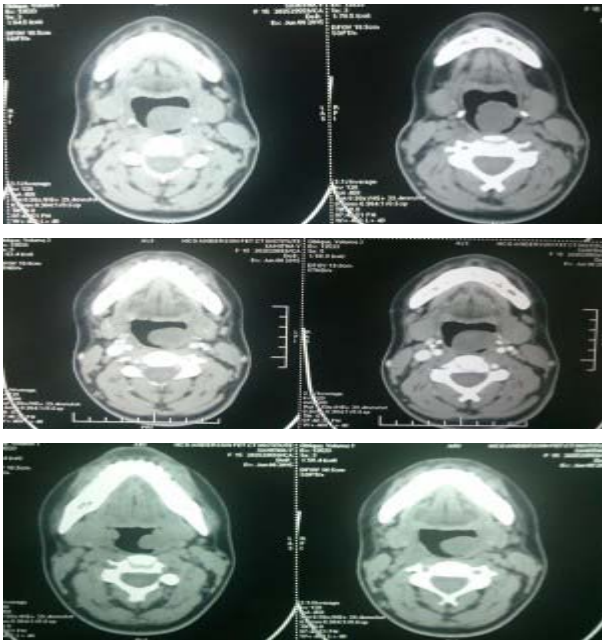
16 year old girl presented with throat pain for 25 days and difficulty in swallowing for 10 days. She was on tab Eltroxin for hypothyroidism. On Examination, a slough covered pedunculated exophytic mass seen arising from the left lateral pharyngeal wall from the inferior pole of tonsil. Video Laryngoscopy confirmed the findings with the mass seen obstructing the view of the left cord. Right cord was mobile. CECT neck showed a well circumscribed soft tissue lesion (19 x 29 x 33 mm) without calcification extending from left tonsillar fossa into vallecula. Tumour was excised trans-orally in Rose's position. HPE diagnosis was Biphasic Synovial Sarcoma showing intersecting bundles of spindle cells with few foci of groups of cuboidal cells/tubules (epitheloid component). There was mild pleomorphism with numerous mitosis.CD99, CK, Epithelial membrane antigen, CD55, CD57 annd Bcl-2 were positive. Ki-67 was positive in 50%. Case was presented to Tumor Board who suggested Chemotherapy (Doxorubicin and Ifosfamide).

VIDEO LARYNGOSCOPY:

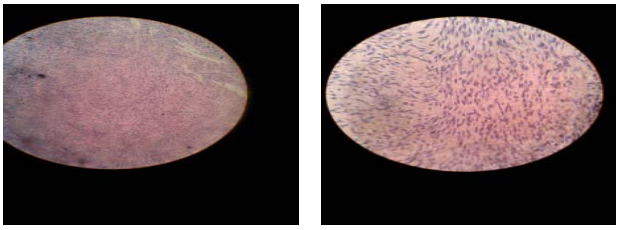


IMAGING:





MICROSCOPY:



Microscopic picture showing intersecting bundles of spindle cells with few foci of groups of cuboidal cells/tubules (epitheloid component)

IMMUNOHISTOCHEMISTRY:

IHC MARKERS	RESULT
CD99	POSITIVE in majority of tumour cells
CK	POSITIVE in morphologically noted tubules
EMA	POSITIVE in many spindle tumour cells, highlights tubules
CD56	POSITIVE in some tumour cells
CD57	POSITIVE in some tumour cells
CD34	NEGATIVE
CD10	NEGATIVE
CK7	NEGATIVE
CK8/18	NEGATIVE
B-100	NEGATIVE
Bcl-2	POSITIVE moderately in majority of tumour cells
KI-67	50%

DISCUSSION:

Synovial sarcoma is an aggressive malignant soft tissue tumor arising predominantly in the lower extremities; only approximately 3–5 % of all cases occur in the head and neck region, and the hypopharynx is the most common site. However, other sites (masticator space, sinonasal cavity, pharynx, parapharyngeal space, tongue, and trachea) may also give rise to Synovial sarcoma (1-5,7- 8) . It has a bimodal age distribution with incidence peaking in the under fives and the adolescents (1, 2, 7-9). In general, males and females are almost equally affected (10,11).

The mutation found most commonly is translocation of SYT gene and a SYT/SSX1 fusion type. Clinically, the symptoms are related to the respiratory and digestive tracts, with dysphagia and dyspnea. Microscopically, SS may be classified into four types: biphasic type, monophasic fibrous type, monophasic epithelial type, and poorly differentiated type (12,13). The diagnosis requires immunohistochemical and cytogenetic examination. By immunohistochemistry, both the epithelial and spindle cell elements of SS show positive immunostaining for vimentin, various cytokeratins, and epithelial membrane antigen (EMA). Synovial sarcoma may also stain for Bcl-2 and CD99. In up to 21 % of tumors, the S-100 protein may be focally expressed (14).

The tumor cells show strong, diffuse, positive nuclear staining for the TLE1 antibody (transducin-like enhancer of split 1)/E , which is a highly sensitive marker of synovial sarcoma. TLE1 is a transcriptional corepressor that binds to a number of transcription factors and plays an important role in the WNT/b-catenin signaling pathway, which is known to be associated with synovial sarcomas. Positive nuclear expression of TLE1 occurs in more than 90 % of synovial sarcoma cases, typically in more than 50 % of the cells . At least 95 % of all SS bear a unique chromosomal translocation, which results in a fusion of the SYT gene on chromosome 18 with either the SSX1 gene or SSX2 gene or, more rarely, the SSX4 gene on the X chromosome. Whether different fusion types influence the outcome of the disease is still controversial. Because these gene fusions are highly specific, their detection with molecular genetics allows the pathologist to render a correct diagnosis. For application in formalin-fixed and paraffinembedded tissue the two molecular biologic methods currently available are conventional and variations of reverse transcription-polymerase chain reaction (RT-PCR) and fluorescence in situ hybridization (FISH). RT-PCR has a higher sensitivity than FISH (94 vs 82 %) and a specificity and positive predictive value of 100 % and a negative predictive value of 80 and 75 % respectively.

The optimal approach to treatment in SS in general and HNSS in particular remains to be determined. The standard treatment for localized disease is surgery with adequate wide excision. Radiotherapy seems to have an established role in improving local control after inadequate surgical resection. The role of post-operative chemotherapy remains unclear. The differences in metastasisfree survival among the various SYT-SSX fusion types could not be attributed to adjuvant chemotherapy. In summary, we reported a case of a primary synovial sarcoma of the tonsil with immunohistochemical studies confirming the same. Synovial sarcoma is a rare malignant soft tissue tumor that rarely involves the tonsil; nevertheless, it should be considered in the differential diagnosis of tumors in this location.

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University Journal of Surgery and Surgical Specialities