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Case Report

CASE REPORT OF SYNOVIAL SARCOMA PRESENTING AS A THIGH SWELLING

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ABSTRACT

Synovial sarcoma (SS) is a rare tumor arising from the mesenchymal lining and commonly arising near proximity of joint. The tumor is difficult to diagnose initially or can be misdiagnosed due to its site and slow-growing potential. This is a malignant tumor with genetic predisposition and has a poor prognosis, if metastasizes to distant organs. The current study tries to describe the clinical, histopathological and immunochemistry findings of synovial sarcoma. A case of SSs presented in our institute for surgery and histopathological examination. Relevant history was taken retrospectively and consent of the patient was also taken. The treatment modality consists of wide excision surgery, chemotherapy, and radiotherapy. In this case, a 35 years old having tumor growth involving thigh area, the excised tumor was sent to our department for histopathological examination. On HPE, monophasic spindle cell tumor was diagnosed with differential diagnosis of SS or malignant peripheral nerve sheath tumor and immunohistochemistry staining was done for further confirmation.

Keywords: Spindle cell, Synovial sarcoma, Thigh, Immunohistochemistry.

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INTRODUCTION

Synovial sarcoma (SS) is a common soft-tissue tumor arising from mesenchymal cell with variable epithelial components. They comprise 5–10% of total soft tissue tumor [1]. The most common site of occurrence is around large joints frequently knee joints and ankle, compromise approximately two-third of total incidences. Its name synovial is a misnomer as it is not arising from synovium and extra articular in presentation, though originating primarily near the joints. SSs arise in close proximation to the tendons, sheath, and bursa. Although it can occur any where in the body but common areas of involvement are such as upper extremity, trunk, head- neck and viscera [heart, lung, pleura and kidney], retroperitoneal and nervous system [2]. SS most commonly affects age group of 10-40 years that is adolescent and young adults, with slightly more male preponderance than females [1,3].

In the early stages of growth, it may be missed out as it is slow-growing with minimal or no signs or symptoms. It gets noticed by the individual when the size increases or due to pain, if local nerves are involved. It is firm to hard in feel and almost fixed to the site, difficult to move. It is deep seated in muscle and may be painful when size is increased. If nerves are involved the growing tumor compresses it and can present as numbness or tingling sensation. It can be misdiagnosed as arthritis, synovitis, or bursitis, due to similar symptoms and site of origin.

No clear cut causal factor has been identified, though genetics may play important role. Exposure to the chemicals such as thorium dioxide, vinyl chloride, or arsenic can be a factor. What happen in SS is translocation t {X; 18) (p11;q11) involving chromosomes X and 18. The common genes involved are SS18 and either SSX1, SSX2, or SSX4 which is specific for this condition and is almost found in 90% of the cases. This is somatic mutation and the changes are not inherited. Its occurrence is sporadic and does not run-in families. Although it is rare, but association with Li Fraumeni syndrome or neurofibromatosis type 1 is seen [4].

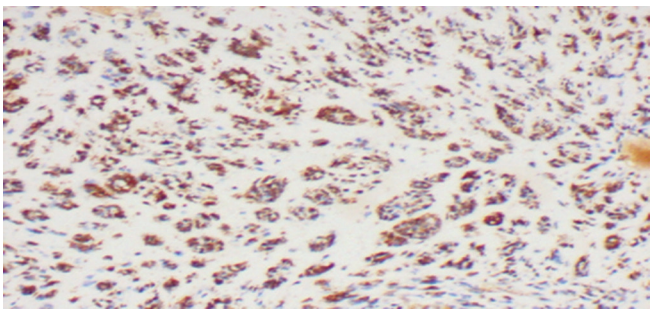
CASE REPORT

A male 35 years old presented to surgery outpatient department of our institute with swelling in the anteromedial aspect of lower end thigh. It is a long-standing swelling, gradually increasing in size for 3–3.5 year. On physical examination, there are no any symptoms except the size as tumor grew large enough about 5.2 × 4 cm. It is fixed to the site with less mobility with no associated pain and numbness. No other lump/lesion identified elsewhere in the body. The patient did not report any past medical/family history. The lesion was evaluated at radiology department, which shows tumor arising from soft0tissue having mild internal vascularity, few necrotic areas, and spotty radiopacities due to focal calcifications reported. The probable diagnosis of mesenchymal tumor, probably sarcoma, was given on USG scan. After that, the lesion was surgically excised in January 2022 in our institute with removal of adjoining normal tissue resection. The excised mass was sent to the department of pathology for histopathological examination. On gross examination, a well-circumscribed globular tissue received, measuring 5.2 × 4 cm. Cut surface is grayish-white in color, with firm to hard in feel. The microscopic examination revealed a cellular monophasic spindle cell tumor with hyperchromatic nuclei and a small intervening stroma. Immunohistochemistry (IHC) was suggested for further confirmation of tumor (Fig. 1).

In IHC, final diagnosis panel was used which contains a number of IHC markers contains DESMIN, SMA, CD34, S-100, Ki-67, CK, MYOGENIN, MYO D1, CD99, SYNAPTOPHYSIN, TLE-2, HMB-45, CD56, EMA, FLI-1, and WT-1. IHC results are suggestive of SS with positivity of Ki-67, CD-56, TLE-1 (Transducin-Like Enhancer 1), and CD-99. The diagnosis of SS among the other soft-tissue sarcoma poses a challenge and may change the line of treatment.

IHC slides –

CD99 – approximately >90% positivity (Immunoreactive, Score 3+).



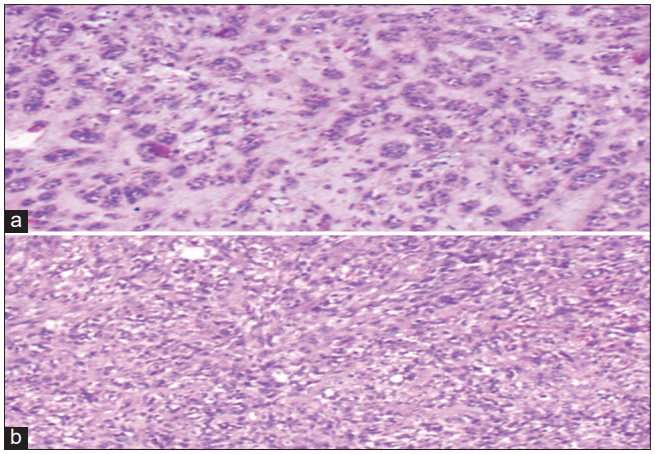
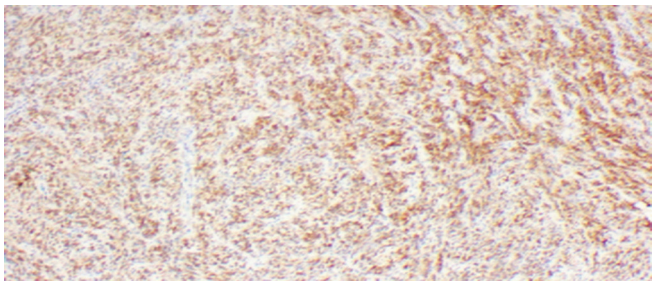


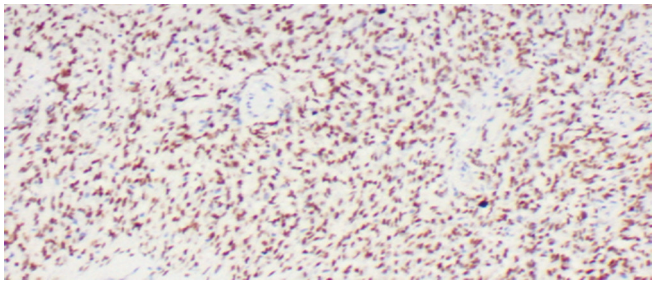
Fig. 1: (a and b) HPE slides (monophasic synovial sarcoma)

CD99 (MIC2): It is mainly used for the diagnosis of Ewings Sarcoma/PNET, but also comes positive in large numbers of tumors including lymphoblastic lymphoma, retinoblastomas, SS, NET, granulosa cell tumor and some variety of rhabdomyosarcomas, and desmoplastic small cell tumors.

CD56- approximately >90% positivity (Immunoreactive, Score 3+).

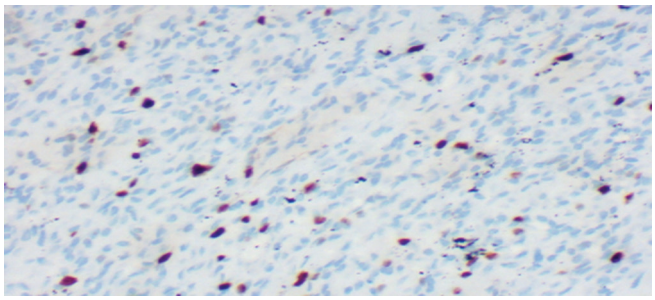


TLE1- approximately >95% positivity (Immunoreactive, Score 4+).



TLE1: (Transducin like enhancer of split 1) overexpression in IHC is highly sensitive and specific biomarker for the diagnosis of SS. It is also positive but weakly in cases of malignant peripheral nerve sheath tumor and solitary fibrous tumor.

Ki67 – Immunoreactive in 10–15% of lesional cells.



Ki67 is a cell proliferation marker that corresponds to a nuclear non-histone protein expressed by the cells in the proliferative phase. Mitotic count and Ki 67 have a directly proportional relationship.

DISCUSSION

The diagnosis of SS is dependent commonly on a histological and immunohistochemical study. A histologically SSs has four subtypes: biphasic, monophasic spindle cell, and poorly differentiated. Biphasic and monophasic fibrous spindle cell variant are commonly seen variant [5]. There is no significant difference among the subtypes. Poorly differentiated SS can be diagnostically challenging as it can occur either in monophasic or biphasic forms. They are located usually in the extremities around large joints most commonly the knee and lower thigh region [6]. Non-aggressive soft-tissue lesions involve intra-articular joint space, commonly villonodular synovitis and osteochondromas; they are the most frequent differential diagnoses for SSs.

Monophasic (epithelial predominant) SS is most important differential diagnosis in this case. SS is a distinctive soft-tissue tumor having epithelial differentiation. IHC and molecular techniques play an important role in the diagnosis. The prognosis for patients with SS of extremities is poor. Systemic chemotherapy is often administered in palliative cases, where surgical resection is not possible [7].

Immunohistochemical analysis and finding the specific chromosomal translocation are necessary, some tumor show immunoreactivity for the cytokeratin (specially CK7 and CK19), EMA, focal positivity for S100, and immunoreactivity for CD99 and CD56 [8]. In CK negative cases, TLE1 is a useful marker for the diagnosis. An important negative IHC marker in SS is CD34 [9].

According to the literature, local recurrence and/or metastatic disease are found in nearly 80% of patients. Several factors have been associated with a higher recurrence risk and poor prognosis. These factors include older age, larger tumor size (> 5 cm), truncal location or proximal tumors in the limbs, male sex, biphasic tumor, bone or neurovascular invasion, incomplete excision on pathological examination, p53 overexpression, high proliferative index, and, more recently, specific SYT-SSX fusion types [10]. In this case, the patient presented with only three of these risk factors. The most common site for metastasis is the lung, and lymph node involvement in 27% of patients, that was absent in this case. Follow-up of this case can guide us to better management and track progression.

AUTHORS' CONTRIBUTIONS

Dr Binay Kumar was involved in collecting, diagnosis, case study analysis, reviewing, and editing the data. Dr Nidhi Prasad was involved in case study analysis, drafting, and reviewing the manuscript.

ETHICAL APPROVAL

Not applicable in this study as patient identity is not revealed and compromised.

CONFLICTS OF INTEREST

None.

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REFERENCES

1. Siegel HJ, Sessions W, Casillas MA Jr., Said-Al-Naief N, Lander PH, Lopez-Ben R. Synovial sarcoma: Clinicopathologic features, treatment, and prognosis. Orthopedics 2007;30:1020-5; quiz 1026-7. doi: 10.3928/01477447-20071201-15, PMID 18198773
2. Steinstraesser L, Agarwal R, Stricker I, Steinau HU, Al-Benna S. Biphasic synovial sarcoma of the extremity: Quadruple approach of

isolated limb perfusion, surgical ablation, adipofascial perforator flap and radiation to avoid amputation. Case Rep Oncol 2011;4:222-8. doi: 10.1159/000327845, PMID 21516237

3. Sultan I, Rodriguez□Galindo C, Saab R, Yasir S, Casanova M, Ferrari A. Comparing children and adults with synovial sarcoma in the surveillance, epidemiology, and end results program, 1983 to 2005: An analysis of 1268 patients. Cancer 2009;115:3537-47. doi: 10.1002/cncr.24424, PMID 19514087
4. Correa H. Li-Fraumeni syndrome. J Pediatr Genet 2016;5:84-8. doi: 10.1055/s-0036-1579759. PMID 27617148, PMCID PMC4918696
5. Matthew R, Lindberg DL, Gardner JM, Cassarino DS, Stallings-Archer K. Diagnostic Pathology: Soft Tissue Tumors. Canada: Elsevier; 2016.
6. Rajwanshi A, Srinivas R, Upasana G. Malignant small round cell tumors. J Cytol 2009;26:1-10. doi: 10.4103/0970-9371.54861, PMID 21938141
7. Eilber FC, Dry SM. Diagnosis and management of synovial sarcoma. J Surg Oncol 2008;97:314-20. doi: 10.1002/jso.20974, PMID 18286474
8. Ruggiero A. Synovial sarcoma. Orphanet Encyclopedia March. 2004.
9. Mirzaian E, Tavangar SM, Montazeri S, Yeganeh FE. Biphasic epithelial predominant synovial sarcoma presenting as painful thigh mass. Iran J Pathol 2019;14:261-4. doi: 10.30699/ijp.2019.90673.1865, PMID 31583005
10. Wisanuyotin T, Radapat K, Sirichativapee W, Paholpak P, Kosuwon W, Sumnanoont C, *et al*. Prognostic factors and clinical outcomes in synovial sarcoma of the extremities. Asia Pac J Clin Oncol 2013;9:80-5. doi: 10.1111/j.1743-7563.2012.01563.x, PMID 22898085