

**Mesentric Synovial sarcoma: A Rare Case Report**Dnyaneshwar S. Jadhav<sup>1</sup>, Balaji D. Baste<sup>2\*</sup><sup>1</sup>Associate Professor, Department of Pathology, SRTR Government Medical College & Hospital, Ambajogai, Beed, Maharashtra, India<sup>2</sup>Assistant Professor, Department of Pathology, Seth G.S. Medical College and K.E.M. Hospital, Mumbai, Maharashtra India**\*Corresponding author**

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**Abstract:** Synovial sarcoma (SS) is a rare malignant mesenchymal tumor mainly arising in the peri-articular tissue in young adults. There are few cases reported in other areas. A 26-year-old woman presented with a palpable abdominal mass. USG abdomen showed evidence of an ill defined, multi lobulated mass lesion seen in the peritoneal cavity on left side. Based on histopathological and immunohistochemical (IHC) examination a diagnosis of Synovial sarcoma was given. A synovial sarcoma should always be considered in the differential diagnosis, and immunohistochemistry is an important adjuvant tool in this situation. We report a rare case of primary synovial sarcoma arising in the mesentery.

**Keywords:** Synovial sarcoma, Mesentery, Immunohistochemistry.

**INTRODUCTION**

Synovial sarcoma is estimated to constitute 10% or less of all sarcomas[1]. Synovial sarcoma is a distinctive malignant soft tissue neoplasm that commonly occurs in the extremities of young adults. Although frequently associated with joints, tendons and bursal structures, it is now believed that this tumour does not originate from synovial cells as originally postulated. In fact, these tumours show epithelial differentiation and are thought to be derived from pluripotent mesenchymal cells capable of epithelial differentiation [2].

However, it may also arise in unexpected sites, such as head and neck regions and even the genitourinary tract [3].

Primary intra-abdominal or retroperitoneal SS is a rare condition. Typically, they occur in periarticular locations, with a greater propensity for the lower extremities. In the literature, there have been an increasing number of reports of SS being found in various locations throughout the body. The median age at diagnosis is 35, and there is no predilection for either sex. Unlike other soft tissue histologies, SS has no identifiable etiologic agent or genetic condition that predisposes an individual to develop this malignancy[4].

**CASE REPORT**

A 26 year female was admitted to hospital due to chief complaints of pain in abdomen and vomiting since 1 month. On physical examination, there was palpable lump in the periumbilical region. All routine investigation was within normal range. USG abdomen showed evidence of an ill defined, mixed echogenicity, multi lobulated mass lesion seen in the peritoneal cavity on left side extending from Para umbilical region Up to the aortic bifurcation and touching the anterior

abdominal wall, showing areas of cystic degeneration and necrosis, suggestive of neoplastic mass lesion arising from the mesentery.

CA-125 value measured by Chemiluminescence (CLIA) technology was 232.3 U/ml, because of which clinicians suspected the mass to be ovarian. Non- Contrast computerized tomography(NCCT) Abdomen and pelvis study shows a heterogenous density solid cystic right adnexal mass, another large heterogenous density soft tissue mass within peritoneal cavity, retroperitoneal and mesenteric lymphadenopathy and gross ascites, likely etiology being GIST, Peritoneal sarcoma, Carcinoma ovary with peritoneal deposits.

Intraoperative findings include a large, solid, mesenteric mass (11x10x7cm) arising from the mesentery of the small bowel starting from 15cm of duodeno -jejunal junction and distally upto 30 cm away from illeocolic junction. Mesenteric mass was attached to the small bowel and mass had a hemorrhagic clot within it.

Grossly, Fig 1(A) & (B), we received an intestinal segment of 70 cm length with an attached

mesenteric mass of size 11x10x5cm. It was ill defined, globular, and firm in consistency with hemorrhagic and necrotic areas in it on cut section. A single lymph node of 0.5 cm was also identified.

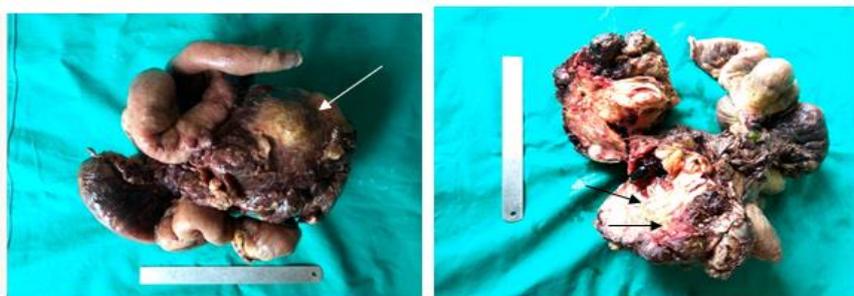
On microscopic examination, tumor tissue was composed of oval to spindle cells with hyperchromatic spindle shaped nuclei with prominent nucleoli. The tumor cells were arranged in fascicular pattern with formation of hypocellular areas. Section also shows evidence of mitosis, focal necrotic areas and haemorrhagic areas. The surgical margins were free of tumour tissue and lymph node showed evidence of reactive lymphadenitis. Malignant Spindle Cell tumor, Leiomyosarcoma and Malignant GIST were the differential diagnosis based on histopathological findings. Immunohistochemistry was advised for confirmation and further typing.

On immunohistochemical analysis, Fig 3(A) & (B), there was positivity for vimentin, CD 99, bcl-2,

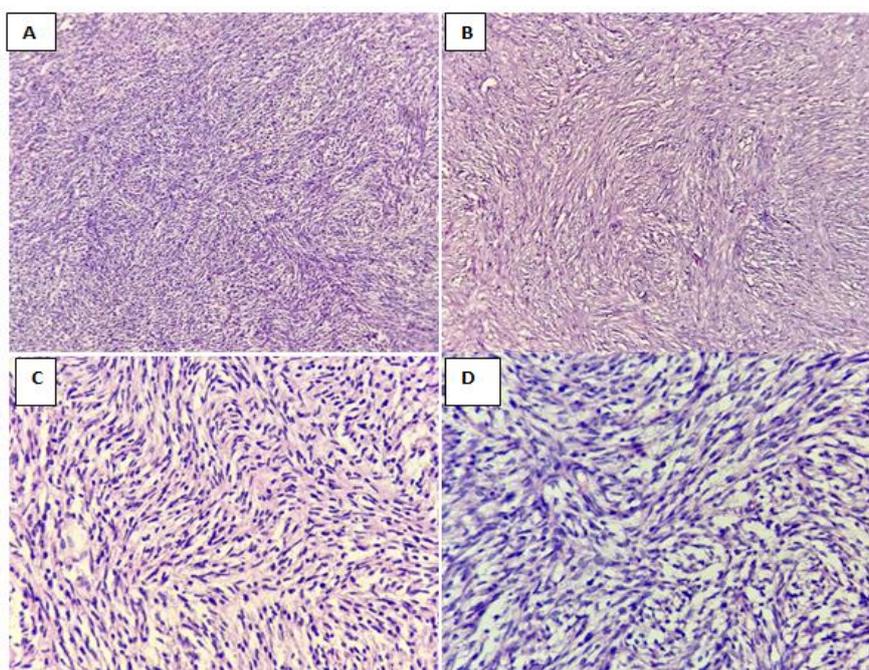
along with focal positivity for pancytokeratin. The cells were negative for Calretinin, CD45, chromogranin, SMA, CD117, CD34, Desmin and S-100P. The study thus excludes the possibility of leiomyosarcoma, gastrointestinal stromal tumour, and usual spindle cell neoplasm in mesentery.

Since tumour cells were vimentin, bcl2 and CD99 positive and with spaces lined by CK positive cells, it points towards the diagnosis of synovial sarcoma. SYT-SSX1 and SYT-SSX-2 gene testing was suggested for confirmation. However it could not be carried out because of unavailability of resources for the same.

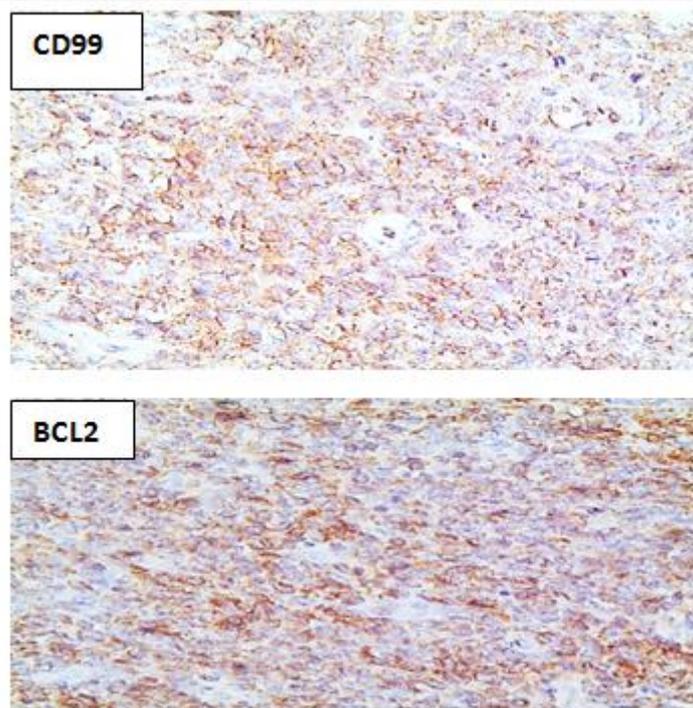
In this case, local recurrence in the retroperitoneal region and bladder with massive ascites developed 2 months after complete resection of the primary tumor. Descending aggression was noted and supposed to be the main reason causing death in this patient.



**Fig-1(A) & (B): Gross ill-defined mass globular and firm in consistency (arrow) with hemorrhagic and necrotic areas on cut section (arrows)**



**Fig-2(A) (B) (C) (D): Tumor tissue was composed of oval to spindle cells with hyperchromatic spindle shaped nuclei with prominent nucleoli. The tumor cells were arranged in fascicular pattern with formation of hypocellular areas. Sections also showed evidence of mitosis, focal necrotic areas and haemorrhagic areas.**



**Fig-3: (A) & (B): Immunohistochemistry: showing membranous staining by CD 99 and nuclear staining by Bcl2**

## DISCUSSION

Synovial sarcoma is a rare soft tissue malignant tumor. Despite its name, SS does not necessarily arise from synovial or soft tissue and has been reported to develop at other sites, such as the kidneys, lungs and pleura. SS most commonly develops in the extremities, particularly in the knee, but primary SS has rarely been reported, with only 8 cases published in the literature to date[5].

There are three main histologic subtypes of SS: biphasic, monophasic and poorly differentiated. The biphasic type represents 20-30% of lesions and has both mesenchymal spindle cell components and an obvious epithelial component usually forming glands. The monophasic type is the most common (50%-60%), in which the spindle cell component predominates. Poorly differentiated SS are epithelioid in morphology and have high mitotic activity. This type has the poorest prognosis [6].

Intra-abdominal SS is rarely found. Immunohistochemistry is useful for diagnosing and distinguishing SS from other malignancies. Most SS are focally positive for cytokeratin and EMA. It has recently been suggested that EMA, cytokeratin AE1/AE3, and E-Cadherin, in combination with CD34 negativity, are the most useful and sensitive protein biomarkers for diagnosing monophasic fibrous pattern, and also good for diagnosing the scantily differentiated synovial sarcoma. In addition, bcl-2 and vimentin were reported as being diffusely expressed in spindle cells of synovial sarcoma[7].

Recent studies have shown the usefulness of cytogenetic and molecular techniques as diagnostic adjuncts for distinguishing SS from other morphologically similar spindle cell sarcomas [8].

When available, the presence of a SYT-SSX1 or SYT-SSX2 fusion transcript resulting from a chromosome translocation t(X;18) (p11.2;q11.2) is a specific finding. Apart from its diagnostic value, the presence of this translocation fusion type has proved to be the single most significant prognostic factor in a multivariate analysis in SS patients. However, a European retrospective analysis found that the most important factor in determining the prognosis of the patient is the histological grade rather than the SYT-SSX fusion type. Patient's age, tumor location, differentiation grade, mitotic activity, neurovascular invasion and SYT-SSX fusion type have variably proved to be independent predictors of survival. Synovial sarcoma has a propensity to local recurrence, particularly when excised without a margin of macroscopically normal tissue [9].

We made the diagnosis in our patient's case based on the combined histological and Immunohistochemical profiles which were highly characteristic of monophasic synovial sarcoma.

## CONCLUSION

In conclusion, synovial sarcoma is a high-grade malignancy with highly metastatic potential; correct and early diagnosis of synovial sarcoma may impact treatment. Our case report emphasizes the importance of accurate histopathological diagnosis, and

use of appropriate immunohistochemical markers in the diagnosis of this unusual tumour in an unusual site.

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