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MONOPHASIC SYNOVIAL SARCOMA - A RARE PRESENTATION

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Abstract : we present a large heterogenously enhancing, well defined, lobulated multilocular cystic lesion seen involving abdominal wall. Biopsy followed by total resection of the lesion showed a spindle cell sarcoma with hemangiopericytomatous pattern, which was consistent with monophasic synovial sarcoma on histology and immunohistochemistry. The unusual clinical presentation, radiology, pathology and differential diagnosis will be discussed in detail.

Keyword :hemangiopericytoma-like, monophasic, synovial, sarcoma

INTRODUCTION

Synovial cell sarcoma is one of the most common soft-tissue tumors in adolescents and young patients, with approximately one third of cases occurring in the first two decades of life. Mean age of patients at diagnosis is approximately 30 years. The origin of synovial cell sarcoma is unclear. The basis for the name synovial cell sarcoma is the similarity between cells of this tumor and primitive synoviocytes. A neurologic origin for this sarcoma has been suggested. Synovial cell sarcoma is characterized by a specific chromosomal translocation, t(X;18) (p11;q11), noted in more than 90% of cases[1]. This fusion gene is called the *SYT-SSX1*, *SYT-SSX2*, or *SYT-SSX4*. These terms correspond to a fusion of the *SYT* gene (chromosome 18) with the *SSX* gene (chromosome X). Males are more commonly affected than females[2]. Biphasic tumors express the SYT-SSX1 transcript, whereas monophasic tumors with only a spindle cell component may express either transcript[3].

PRESENTATION

A 71-year-old male presented with a 2 month history of mass and non-radiating, dull-aching upper abdominal pain, without associated vomiting or altered bowel habit. Local examination revealed 20x15 cm mass in left hypochondrium, firm, not mobile, not tender. No organomegaly noted. CT abdomen showed a large complex 17x13x15 cm solid cystic multilocular mass in left subcoastal region involving abdominal wall muscles mimicking abscess[fig.1]. The lesion biopsied and sent for histopathology.

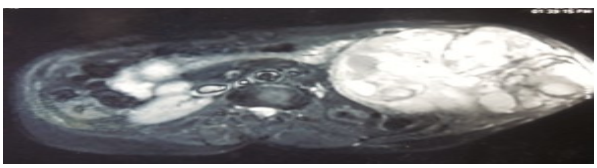

Fig.1 : CT showing multilocular cystic mass

OBSERVATION

Grossly, the tumor was well circumscribed tan to gray. The cut surface was soft to firm. Cyst formation was prominent. This case is presented for its rare presentation which was predominantly cystic with cyst containing necrotic slough, pus and blood as is evident from its gross appearance which mimics an abscess[fig.2]. Calcification can be grossly apparent and may be detected radiographically but was absent in our case. H and E stained sections showed a capsulated malignant spindle cell tumor with hemangiopericytomatous (HPC) pattern of vessels[fig.3,4]. Spindle cells were arranged in fascicular pattern with cells having indistinct cytoplasm, hyperchromatic nucleus and few mitoses, interspersed with numerous capillaries, along with large areas of necrosis and haemorrhage. we arrived at diagnosis of malignant spindle cell neoplasm probably synovial sarcoma or fibrosarcoma and proceeded to immunohistochemistry for confirmation. IHC showed bcl-2 positivity in 90-100% of tumor cells[fig.5]. EMA [fig.6] showed focal mild positivity and Vimentin[fig.7] was positive in 100% of tumor cells and CD99 showed focal positivity. S100, CD34, SMA, c-KIT was negative in tumor cells. Thus H&E along with IHC confirmed the diagnosis of synovial sarcoma.

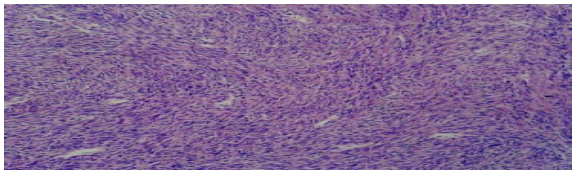

Fig.2 : Central necrotic areas mimicking abscess Fig.3 : HPC like pattern

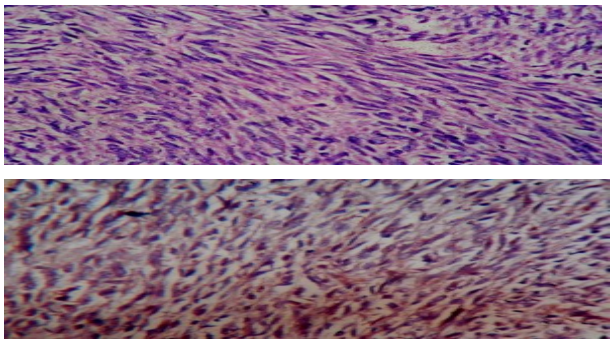

Fig.4 : Intersecting bundles of malignant spindle cells
Fig.5 : BCL-2 diffuse strong positivity

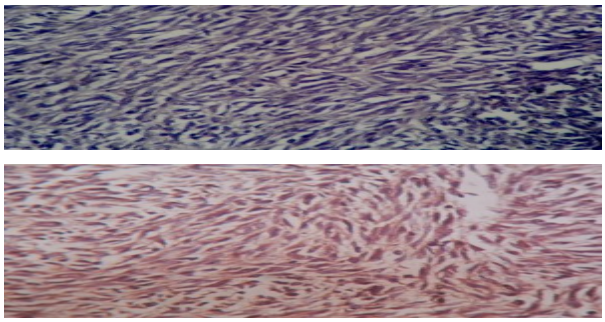

Fig.6 : EMA focal mild positivity
Fig.7 : Vimentin diffuse strong positivity

DISCUSSION

Synovial sarcoma was a well-defined clinical and morphological entity that was originally described by Simon in 1865 and was so named in 1934 by Sabrazes[4]. The term “synovial sarcoma” is however a misnomer as the tumor cells do not share the same immune-histochemical and ultrastructural features of the normal synovium. The cellular origin of this tumor is probably considered as neural crest derived cells[5]. This tumor accounts for 5–10% of the soft tissue sarcoma[6]. The majority of synovial sarcomas occur in the extremities with a predilection for the lower extremities. Less frequently, occurence in the head and neck, abdominal wall, pleura, lung, mediastinum and in almost all anatomical sites has been noted. The lesions often grow close to joints, particularly the knee, however the intra-articular tissues are only rarely involved. They vary in size and are often deep-seated. Slow-growing tumors tend to be well- circumscribed, while rapidly growing tumors tend to be more variegated. Immunohistochemically, SS are positive for cytokeratin (either AE1/AE3 or CAM5.2), epithelial membrane antigen, vimentin, bcl-2, CD99 and calponin. Keratin markers and EMA has been reported to react in 60-70% of the cases of monophasic SS while immunoreactivity for S100 protein has been reported in about 30% of the cases. *TLE1*, a gene, related to WnT pathway and this has been found to be consistently overexpressed in SS[8]. Recent studies have shown the usefulness of cytogenetic and molecular techniques as gold-standard diagnostic tools for SS[9].

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