

MONOPHASIC SYNOVIAL SARCOMA-A RARE CASE REPORT IN A 17 YEAR OLD FEMALE

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ABSTRACT

We present this rare case of ours in a 17 year old girl who presented with pain and swelling of left knee joint since five years. She noticed a recent rapid increase in size for the past one month. Clinical and Radiological examination revealed a left soft tissue lesion on the medial side of knee joint of size 10x6cm with a possible diagnosis of Hemangioma and Myositis ossificans. Surgical resection of cystic swelling was done. The histological and immunohistochemical findings were consistent with final diagnosis of synovial sarcoma monophasic type.

Key Words: Young Adults, Soft Tissue Tumour, Knee Joint, Monophasic Synovial Sarcoma

INTRODUCTION

Synovial sarcoma is a rare soft tissue malignant neoplasm arising in deep soft tissue of extremities, especially around the knee and adjacent to large joints or tendon sheaths.. Synovial sarcoma is a misnomer because the tumour does not arise from the synovium Synovial Sarcoma accounts for 7to 10% of soft tissue sarcomas (Rajwanshi et al., 2009). They are reported from birth to 89 years but occur mainly in young adults (Ruggiero, 2004) and more commonly in males; 90% of cases occur before 50,in between 15 and 35 years.

CASE REPORT

A 17 year old girl presented to surgical clinic with pain and swelling on left medial side of knee joint. Patient was apparently asymptomatic 5years back then she met with an accident and developed a swelling on medial of the left knee joint which was initially small in size which progressed to the present size with rapid increase in one month. Past history was nil significant. Clinical examination revealed no organomegaly and no lymphadenopathy. There was no loss of appetite or no loss of weight. Local examination swelling of 10cm x 6cm present over left medial side of knee joint , oval in shape, firm in consistency with smooth surface well defined margins , movable in both horizontal and vertical planes , in subcutaneous plane, skin over swelling was normal and was tender to touch.

Radiological findings

X-ray Examination of left knee joint showed a soft tissue mass lesion of 9x6x4 cm with cystic degeneration adjacent tibia showed rarefaction changes.

Magnetic Resonance Imaging T1 showed a large well defined mixed signal intensity lesion with peripheral zone of nodular rim hypo intensity on the medial aspect of the knee joint at the level of proximal tibia in the subcutaneous plane noted on the GRE sequences(likely due to ossification. The most peripheries of the lesion shows layering of different zones.T1 sequences shows areas of chronic hemorrhage with fluid signal intensity in the form of multi-locular changes.

Doppler test screening of the lesion superiorly shows increased vascularity with layering of calcification.

Differential diagnostic -possibilities of

1. Myositis ossificans
2. Extra skeletal osteosarcoma to be considered.

Synovial fluid analysis cytospin cytosmear shows mostly blood elements with few macrophages. Other laboratory investigations were within normal limits. Excision biopsy of the swelling was done and sent to pathology department.

Gross pathology: we received grey white soft tissue mass measuring 9x8x3cm, relatively well circumscribed lobulated. Firm in consistency, Cut surface showed fleshy variegated appearance with grey white and grey brown areas.

Microscopic examination:

The histology demonstrated tumour tissue arranged in diffuse sheets, fascicles, swirling and pertheliomatous pattern. Individual cells were plump spindle shaped with scant to moderate amounts of eosinophilic cytoplasm, spindle nuclei with dispersed chromatin, Inclusions and nucleoli in some were noted. Mitotic activity was brisk with 3-

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4/HPF. Areas of cystic degeneration, necrosis, calcification and hemorrhages were also seen focally. Diagnosis suggesting soft tissue sarcoma- monophasic synovial sarcoma.



Figure 1: X-ray AP view showing soft tissue lesion on medial side of knee joint



Figure 2: Gross specimen showing well circumscribed lobulated soft tissue mass cut surface grey white to grey brown

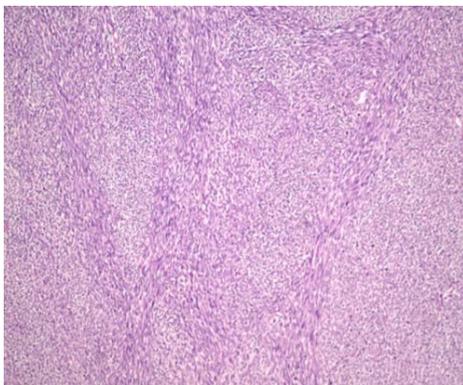


Figure 3: H&E: tumour tissue arranged in fascicles and bundles 10x

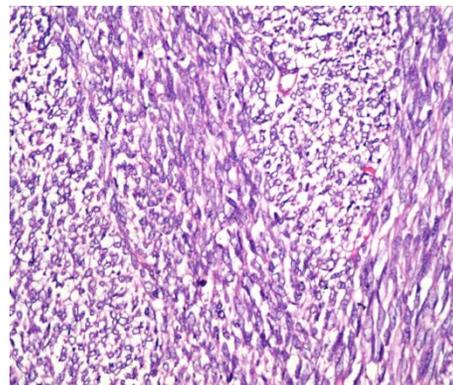


Figure 4: H&E individual cell are spindle shaped with elongated vesicular nuclei. 40x

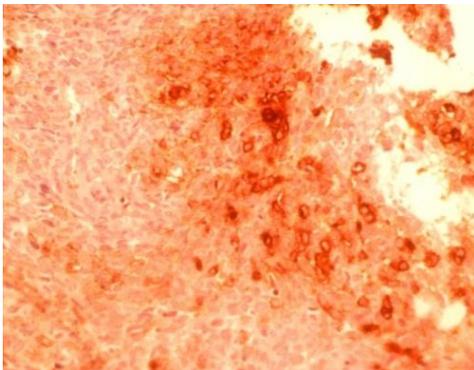


Figure 5: 40x, EMA(E29) –focal membrane positivity

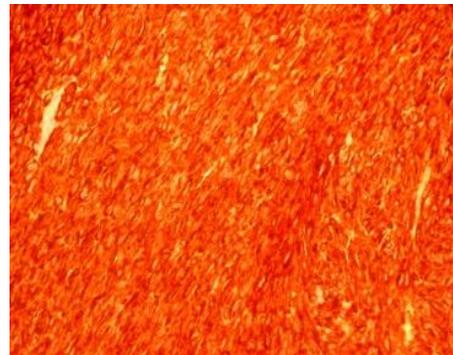


Figure 6: 40X, bcl2(100)-strong nuclear and cytoplasmic positivity

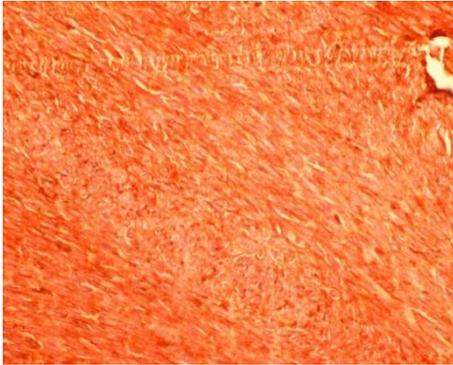


Figure 7: 40X, Cd99(HO36.1.1)-diffuse membrane positivity.

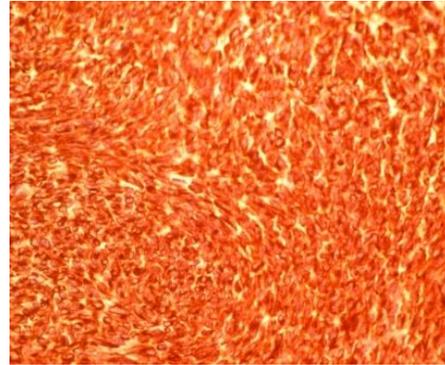


Fig8: 40X, Vimentin(V9)- diffuse cytoplasmic positivity

Immunohistochemical workup confirmed the diagnosis Cd99 (HO36.1.1)-diffuse membrane positivity, EMA (E29)-focal membrane positivity, bcl2(100)-strong nuclear and cytoplasmic positivity, Vimentin(V9)-diffuse cytoplasmic positivity (Fig. 1-8).

DISCUSSION

Synovial sarcomas account for about 7-10% of soft tissue sarcomas. Synovial sarcoma is a mesenchymal spindle cell tumour which displays variable epithelial differentiation, including glandular formation and has a specific chromosomal translocation $t(X;18)(p11;q11)$ (Afif et al., 2006). There is mild male predominance; the sex ratio is 1:5. It affects preferentially young patients.

Clinically the features may be non-specific and does not distinguish it from other sarcomas. They often present as slow growing well circumscribed mass that mimics benign pathology.

Grossly tumours are round or multilobular poorly or well circumscribed. They may be either unencapsulated or surrounded by fibrous capsule (Suster & Moran, 2005). On cut section they are grey yellow and exhibit variegated appearance with cyst formation, hemorrhage and necrosis. Gross calcification may be evident (Gaertner et al., 1996).

Synovial sarcoma is composed of two morphologically different types of cells: epithelial cells, resembling those of carcinoma, and fibrosarcoma-like spindle cells. Depending on the relative prominence of the two cellular elements and the degree of differentiation, synovial sarcomas form a continuous morphologic spectrum and can be broadly classified into the (I) biphasic type, with distinct epithelial and spindle cell components in varying proportions; (II) monophasic fibrous type; (III) rare monophasic epithelial type and (IV) poorly differentiated (round cell) type (Synovial, 1995). Almost all morphological subtypes are characterized by a specific $t(X;18)(p11.2; q11.2)$ chromosomal translocation.

Like other soft tissue sarcomas, synovial sarcoma's diagnosis is difficult to establish purely on the basis of histological appearance. It is even difficult in some cases without an obvious biphasic differentiation. Hence, immunohistochemical studies must be completed showing neoplastic cells diffusely immunoreactive to CK, EMA, Vimentin, Bcl-2, Actin and CD99 with focal immunoreactivity for S-100 protein and are negative for CD34 and Desmin (Pelms et al., 2002).

The monophasic fibrous synovial sarcoma (such our case) may resemble a number of other spindle cell neoplasm's including fibrosarcoma, leiomyosarcoma, MPNST, hemangiopericytoma, and spindle cell carcinoma. In fact, the spindle cells of fibrosarcoma appear in a bands of interlaced arrangement, mitotic figures are common, and epithelial markers are negative. The spindle cells of leiomyosarcoma have a dark eosinophilic cytoplasm and Smooth Muscle Actin or Desmin are positive. MPNST is of neural origin, so the spindle cells are more wave-shaped, and one end of the nuclei is bulged, and S-100 is positive but epithelial markers negative. Hemangiopericytomas also need to be differentiated from synovial sarcoma. These tumors have vascular changes in all of the tumor area. Cells are polygonal in shape, CD34 positive and negative for epithelial markers (Bui-Mansfield et al., 2002).

Treatment of choice of synovial sarcoma is multimodal combination of wide-to radical resection, radiation therapy and adjuvant chemotherapy following resection and since synovial sarcoma is known to recur, a careful follow up is mandatory (Oquadnoui et al., 2011; Fatimi and Saleem, 2009). Synovial sarcomas metastasize to bone, liver, skin, the central nervous system, and even breast tissue (Banerjee et al., 2004; Zeren et al., 1995).

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Prognosis is related to the disease stage and is usually poor. Young age, Her-2 expression, complete resection with clear surgical margins and response to first line chemotherapy were found as good prognostic indicators in different studies (Sápi et al., 2005; Régnard et al., 1999). On the other hand, adverse prognostic factors for synovial sarcoma include male gender, truncal as opposed to distal tumor location, lesions larger than 5 cm, high histologic grade (based on the mitotic rate and tumor necrosis), neurovascular invasion, aneuploidy, poor histological differentiation and local recurrence (Bergh et al., 1999; El-Naggar et al., 1990).

CONCLUSION

Synovial sarcoma is rare malignant mesenchymal spindle cell tumour characterized by variable epithelial differentiation. This case is a rare form monophasic type presenting in a young female. It is important to differentiate it from other soft tissue malignancies as clinical presentation is similar in all of them and also the treatment modality differs in each of them. Follow up is advised in these cases.

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