Case Report

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Monophasic synovial sarcoma: a case report and literature review

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ABSTRACT

Synovial sarcoma is an aggressive soft tissue tumor which accounts for 7% - 8% of all human malignant sarcomas. Although this tumor generally affects adults, about 30% of reported cases occur in children and adolescents. Synovial sarcoma is the most common malignant non-rhabdomyosarcomatous soft tissue sarcoma in children and adolescents. We report a case of synovial sarcoma presented with pain and swelling in the left knee. MRI of left knee shows $25 \times 54 \times 56$ mm soft tissue lesion in infrapatellar bursa. FNAC left knee swelling reported malignant spindle cell tumor. The mass was completely resected in a surgical procedure. In pathological examination of the mass, monomorphic fibroblastic synovial sarcoma was reported. Patient was treated with Ifosfamide and Adriamycin.

Keywords: Monophasic, Synovial sarcoma, Paediatric tumor

INTRODUCTION

Synovial sarcoma (SS) is a rare malignant neoplasm seen in capsules of joints, articular tendons and one of the most common malignant soft tissue sarcomas in children and adolescents. They form 5.6% to 10% of all soft tissue malignancies after malignant fibrous histiocytoma, liposarcoma and rhabdomyosarcoma. ^{3,4}

Controversy still exists at its synovial tissue of origin, as it is now universally accepted that it originates from undifferentiated or pluripotent mesenchymal cells both epithelial and mesenchymal differentiation capacity.⁵ Synovial sarcoma appears in third decade in young adults with incidence twice common in males than females.³

CASE REPORT

A 14 year old boy presented in paediatric outdoor with complain of pain in the left knee for one year and a nodular swelling of about 3-4cm in size present over left knee for 5 months which gradually increased in size over

this period. There was a history of trauma while playing at school. Patient had complained of pain in left knee and difficulty during walking. On examination a 3×3.5 cm growth was present over left knee. Growth was firm in consistency; warm, tender, non-reducible and non-fluctuant. Growth was immovable, fixed to the overlying skin and underlying structures. Plain radiograph of knee showed a soft tissue mass with no calcification. On MRI a 24×54×56 mm soft tissue lesion in infrapatellar bursa was detected. A provisional diagnosis of malignant spindle cell tumor on FNAC was made. The patient underwent debulking surgical removal and tissue sent for histopathological examination.

Pathologic findings

Gross examination showed a grey white irregular soft tissue mass measuring $4\times3\times3$ cm with skin attached on one side. Skin flap measured 3.5×2 cm. Outer surface of mass was nodular and irregular. Cut surface was grey yellow with haemorrhagic and necrotic areas. A satellite nodule measuring 1×0.5 cm was present near the tumour

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mass (Figure 1). Tissue was taken, processed and stained by H&E.



Figure 1: Excised soft tissue mass which was received for histopathological examination.

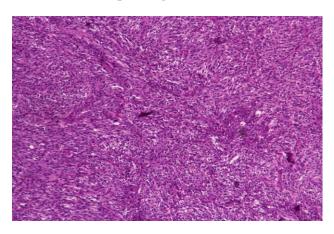


Figure 2: Haematoxylin and eosin stained section showed hypercellular, monomorphic spindle cells (H&E 10X).

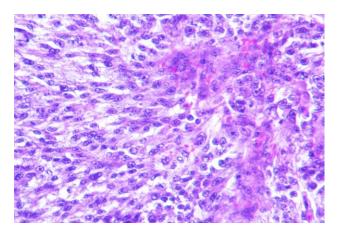


Figure 3: Haematoxylin and eosin stained section showed small, uniform spindle cells, high nuclear cytoplasmic ratio with fine stippled chromatin (H&E 40X).

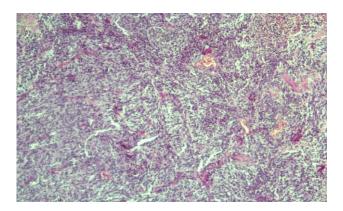


Figure 4: Haematoxylin and eosin stained section showed a branching hemangiopericytoma like vascular pattern (H&E 10X).

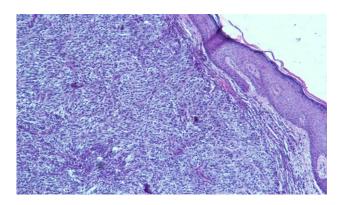


Figure 5: Haematoxylin and Eosin stained section showed tumour cells extend up to epidermis (H&E 10X).

Microscopic examination - Haematoxylin and Eosin stained sections showed a spindle cell neoplasm (Figure 2) made up of small, uniform spindle cells with a high nuclear-cytoplasmic ratio, and finely stippled chromatin (Figure 3). The neoplastic cells were closely packed, growing in short interlacing and intersecting fascicles around a branching, "hemangiopericytoma-like vasculature" (Figure 4). The mitotic rate was 1-2 mitoses/10HPFs. The tumor showed no glandular differentiation. Necrosis was not seen. These tumor cells also extend up to epidermis (Figure 5).

DISCUSSION

Synovial sarcoma is a malignant tumor affecting more often joints of lower extremities than other bones. The knee is most frequent tumor location¹. It often grows close to the joints, tendon sheaths, and bursae but it is extremely rare for it to invade the joint space and synovial membrane, with which it is probably unrelated. The tumor presents in most cases as a painful and tender swelling in the region of a big joint as well as it was the case with our patient who presented with complains of pain and swelling in and around knee joint.

Synovial sarcoma can occur in every age but most commonly in young adults and children with male preponderance². Our patient is also 14 year old male child.

Plain radiographs generally do not provide much information and in about 15% to 20% of cases, underlying bone reaction (such as periosteal reaction, superficial bone erosion, or invasion) or multiple small radio-opacities in form of focal calcification can be seen in X-rays.^{3,4} Radiographic findings of our patient was almost normal except for soft tissue mass.

MRI is the modality of choice in the evaluation of softtissue tumors ^{5,6} allowing examination of the mass in the axial, sagittal, and coronal planes. Although the image does not provide a histological diagnosis, it shows features that allow differentiation between benign and malignant lesions.

The diagnosis is confirmed by excisional biopsy. Grossly it tends to be well circumscribed, firm, and greyish pink. Focal calcification is frequent and may be detected radiographically. Microscopically; classic form of synovial sarcoma is biphasic tumor composed of epithelial and sarcomatous components. The epithelial areas appear in the form of gland like spaces lined by cuboidal (synovial like) or columnar cells. The sarcomatous component is made up of spindle cells with fibroblast like appearance. It is hypercellular with monotonous appearance, plump nuclei, focally whorled pattern, distinct lobulation, hemangiopericytoma like areas. Hyalinization, calcification and osseous metaplasia may be present. When the calcification is heavy, the term calcifying synovial sarcoma has been used.

Monophasic synovial sarcoma is composed of only one of the two components. In the large majority of cases, this applies to the spindle cell sarcomatous component, which is easily misdiagnosed as fibro sarcoma, hemangiopericytoma or some other spindle cell neoplasm by the unwary. ^{9,10}

Histopathological examination of the excised mass from our patient revealed cellular lesion composed of sheets and fascicles of spindle cells with indistinct cytoplasm, irregular nuclear membrane, mild pleomorphism, open chromatin and prominent nucleoli. Foci of necrosis, haemorrhages were seen but there was no evidence of vascular emboli. These features were suggestive of monophasic synovial sarcoma.

The monophasic variant simulate fibro sarcoma, MPNST, SFT/HPC and mesenchymal chondrosarcoma. However clues to real nature of tumor include the presence of small, oval, overlapping nuclei, the identification of rare clusters of more plump and eosinophilic cells, the lack of nerve like wavy nucleus, the lack of any cartilaginous differentiation. Essentially any highly cellular tumor with aforementioned differential diagnosis occurring on extremity of a young adult ought to be considered a

monophasic synovial sarcoma until it is proven otherwise. 11

The optimal approach to the treatment of synovial sarcoma remains undefined because no prospective clinical trials have compared differing therapeutic approaches. ^{12,13} Complete surgical resection of the primary tumor is the mainstay of treatment. This method was used in the management of our case. Adjuvant radiotherapy to treat microscopic residual disease after surgery provides excellent local control and obviates amputation for most patients with extremity tumors. The role of adjuvant chemotherapy remains controversial, and the small numbers of randomized prospective trials complicate interpretation of results. Most regimens that have shown activity in patients with measurable disease include doxorubicin used with or without an alkylating agent (cyclophosphamide or ifosfamide). Despite the chemo sensitivity of synovial sarcoma, some series have failed to show a significant improvement in survival with the addition of chemotherapy. Chemotherapy is probably beneficial only for those with high-risk features of distant disease recurrence, such as large tumor invasiveness, and high histologic grade.

Synovial sarcoma most commonly metastasizes to the lung (74-81%), lymph nodes (12-23%), and bone (10-20%). 14

The prognosis for 5-year survival ranges from 36% to 76%, and the 10 year survival rate is less (20% to 63%) because of late metastases. Favorable prognostic factors include age of the patient (15 years or younger), less than 5 cm in size, distal extremity involvement, and low tumor stage.

It is 13 months since the surgery and there is no untoward event reported. Patient is doing well and there is no sign of recurrence noted till now.

CONCLUSION

After rhabdomyosarcoma, synovial sarcoma is the most common soft-tissue sarcoma in children. The tumor does not appear to arise from the synovium and has the potential to develop almost anywhere in the body. Prognosis is good when the tumor is smaller than 5 cm, non-invasive, and of low or intermediate histologic grade.

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REFERENCES

- 1. Enzinger FM, Weiss SW. Soft tissue tumors. 2nd ed. St. Louis: Mosby Company, 1988.
- 2. Baptista AM, Camarago OP, Croci AT, Oliveira CR. Synovial sarcoma of the extremities: prognostic factors for 20 nonmetastatic cases and a new

- histologic grading system with prognostic significance. Clinics, Sao Paulo. 2006;61(5):381-6.
- 3. Perez CA. Unusual nonepithelial tumors of head and neck. Principles and practice of radiation oncology. 1998;43:1125-34.
- 4. Weiss SW, Goldblum JR. Malignant Soft Tissue Tumors of Uncertain Type. In Weiss SW, Goldblum JR: Soft Tissue Tumors, 4th ed., St. Louis, Mosby, 2001.
- 5. Miettinen M, Virtanen I. Synovial sarcoma a misnomer. Am J Pathol. 1984;117:18-25.
- Bos GD, Esther RJ. Foot tumors: diagnosis and treatment. J Amer Acad Orthop Surg. 2002;10:259-70.
- 7. Machen SK, Easley KA, Goldblum JR. Synovial sarcoma of the extremities: a clinicopathologic study of 34 cases, including semi-quantitative analysis of spindled, epithelial, and poorly differentiated areas. Am J Surg Pathol. 1999;23:268-75.
- 8. Varela-Duran J, Enzinger FM. Calcifying synovial sarcoma. A clinicopathologic study of 32 cases. Cancer. 1982;50:345-52.
- 9. Mackenzie DH. Monophasic synovial sarcoma a histological entity? Histopathology. 1977;1:151-7.

- 10. Miettinen M Lehto VP, Virtanen I. Monophasic synovial sarcoma of spindle cell type. Virchow arch. 1983,44:187-99.
- 11. John S, Brooks J. Disorders of soft tissue. In: Sternberg SS, Antonioli DA, Carter D, Eggleston JC. Sternberg's Diagnostic Surgical Pathology.5th ed. Mills, Stacey E. 2010:173-174.
- Miser JS, Triche TJ, Kinsella TJ, Pritchard DJ.
 Other soft tissue sarcomas of childhood. In: Pizzo
 PA, Poplack DG, eds. Principles and practice
 ofpediatric oncology, 3rd ed. Philadelphia:
 Lippincott- Raven. 1997:865-888.
- 13. Brennan MF, Alektiar KM, Maki RG. Sarcomas of the soft tissues and bones. In: DeVita VT Jr, Hellman S, Rosenberg SA, eds. Cancer: principles and practice of oncology, 6th ed. Philadelphia: Lippincott Williams and Wilkins. 2001:1841-1891.
- 14. Cadman NL, Soule EH, Kelly PJ. Synovial sarcoma: an analysis of 134 tumors. Cancer. 1965;18:613-27.

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