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MONOPHASIC SYNOVIAL SARCOMA - A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Introduction:
Synovial sarcoma is a malignant soft tissue tumour. Of all the malignant sarcomas, synovial sarcoma constitutes about 4-8%.

Aim: To highlight the importance of immunohistochemistry in the diagnosis of synovial sarcoma and discuss the various differential diagnosis.

Material and methods: We report a case of monophasic synovial sarcoma in a 42 year old female who presented with right gluteal mass. FNAC was reported as atypical epithelial neoplasm. Excision biopsy was done and was confirmed as synovial sarcoma by histology and immunohistochemistry.

Conclusion: This case is presented to emphasize on the need of immunohistochemistry to differentiate monophasic synovial sarcoma from other similar lesions.

KEYWORDS : synovial, sarcoma, thigh, monophasic.

INTRODUCTION:

Synovial sarcoma is an aggressive malignant soft tissue neoplasm. It is a misnomer. It doesnot arise from or differentiate towards synovium. Synovial sarcoma is a mesenchymal soft tissue tumour with epithelial differentiation including glandular differentiation. SYT-SSX1/2 t(x:18) translocation is specific for synovial sarcoma. Most of them arise in deep soft tissue of the extremities in young adults with a male preponderance. Different histological types includes monophasic, biphasic, poorly differentiated and calcifying synovial sarcoma. We report a case 42 year old female diagnosed as monophasic synovial sarcoma.

CASE HISTORY:

42 year old female presented with history of gradually increasing painless swelling in the right gluteal region since 1 ½ years.

Clinically, 7x6cm firm to hard lump was felt in the right gluteal region. Radiological investigations revealed a soft tissue mass in the right gluteal region measuring, 8.5x6cm in the subcutaneous plane with infiltration of medial aspect of gluteus maximus muscle.

FNAC yielded haemorrhagic material with epithelioid cells displaying moderate atypia. FNAC was reported as atypical epithelial neoplasm.

Grossly, skin covered soft tissue mass was received measuring 8.5x6x5cm with a scar in the measuring 3cm. External surface appeared nodular. Sectioning revealed a firm grey white haemorrhagic tumour measuring 8x5cm. Margins appeared grossly free of tumour.

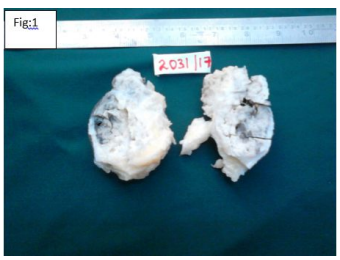

Fig 1: Gross appearance of the tumour- Haemorrhagic firm grey white cut surface

Microscopic examination revealed a tumour arranged in lobules, nests, cords and alveolar pattern composed of small to medium sized polygonal to spindle shaped cells with hyperchromatic nuclei, small nucleoli and moderate amounts of cytoplasm. Occasional mitotic activity is noted. Staging was done, revealed pT2b, FNCLCC tumour grade II.

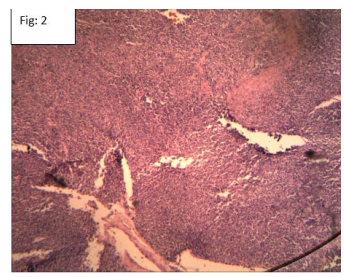

Fig 2: Tumour arranged in lobules (4x)

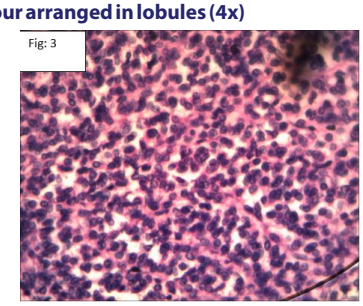

Fig 3: Sheets of polygonal cells with hyperchromatic nuclei (40x)

Immunohistochemistry revealed weak positivity for Cytokeratin and strong positivity for TLE-1 and CD56. Immunohistochemistry for S-100, HMB 45, CD99, myogenin, PAX- 8, EMA, synaptophysin and CD34 were negative.

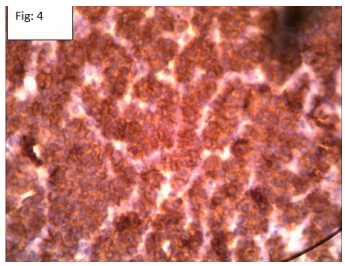

Fig 4: Immunohistochemistry showing strong positivity for CD56

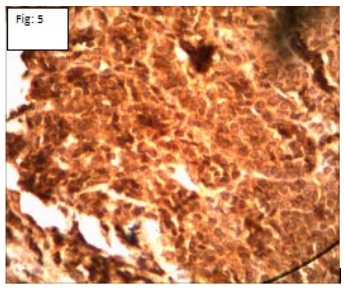

Fig 5: Immunohistochemistry showing positivity for Cytokeratin

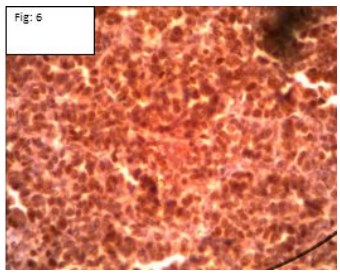

Fig 6: Immunohistochemistry showing strong positivity for TLE -1

DISCUSSION:

Synovial sarcoma is an aggressive soft tissue tumour, occurring in periarticular region. Extremities is the most common site affected. Synovial sarcoma has a male preponderance. Though it can occur at any age, it occurs mostly in young adults. The diagnosis is usually confirmed by histology and molecular translocation for SYT-SSX1/2. The various histological types includes monophasic, biphasic, calcifying and poorly differentiated synovial sarcoma. Classical synovial sarcoma is a biphasic tumour composed of epithelial and spindle cell component. Epithelial component is formed by glands lined by cuboidal or columnar cells. Spindle cell component is usually arranged in fascicles displaying atypia. Hemangiopericytomatous type of vascular pattern can be noted.

Monophasic synovial sarcoma is composed of only either epithelial or spindle cell component. For confirmation of diagnosis, immunohistochemistry is essential. Molecular analysis for specific translocation can also be done.

Differential diagnosis includes malignant peripheral nerve sheath tumour, fibrosarcoma, solitary fibrous tumour, hemangiopericytoma, fibrosarcoma. Synovial sarcoma can metastasize to lung, liver and bones.

The 5 year survival rate is 35-56%.

The prognosis depends on various factors. Good prognostic factors includes age less than 15 years, tumour size less than 5cm, mitotic activity less than 10/10hpf, lower stage and those which originate in distal extremities.

Poorly differentiated synovial sarcoma has a poor prognosis. Presence of SYT-SSX1/2 translocation is associated with a good prognosis.

CONCLUSION:

Monophasic synovial sarcoma must be differentiated from other malignant spindle cell/epithelial cell neoplasms. Differentiation from other neoplasms is possible only by immunohistochemistry and molecular translocation. This case is presented to emphasize on the need of immunohistochemistry to differentiate monophasic synovial sarcoma from other similar lesions.

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