Primary Retroperitoneal Synovial Sarcoma: A Diagnostic Dilemma

Sailuja Maharjan[1], MBBS, MD; Bandana Satyal[1], MBBS, MD; Reena Baidya[1], MBBS, MD; Prakash R Neupane [2], MBBS, MS; Arun Shahi [3], MBBS, MD

[1] Department of Pathology, B & B Hospital, Gwarko, Lalitpur, Nepal

[2] Department of Surgical Oncology, B & B Hospital, Gwarko, Lalitpur, Nepal

[3] Department of Medical Oncology, B & B Hospital, Gwarko, Lalitpur, Nepal

Address of Correspondence:

Sailuja Maharjan, MBBS, MD

Department of Pathology, B & B Hospital, Gwarko, Lalitpur, Nepal,

Email: sailuzaa73@gmail.com

Synovial sarcoma is a rare soft tissue tumor comprising of 1% of all malignancies. It usually involves the extremities in adults in third to fifth decade of life. Retroperitoneal soft tissue sarcoma accounts for only 15% of all soft tissue sarcomas. Here, we report a rare case of primary retroperitoneal synovial sarcoma confirmed by ancillary tests performed on cell block sample.

Keywords: cell block, retroperitoneal, synovial sarcoma.

Synovial sarcoma accounts for approximately 5-10% of all soft tissue sarcomas with extremities being the most common site.[1,2] Other less common sites are head & neck, mediastinum, abdominal wall and retroperitoneum.[3] Primary retroperitoneal synovial sarcoma is an extremely rare entity and has worse prognosis.

Cell block plays a significant role in cytological diagnosis by maintaining architecture which closely resembles that seen in surgical specimens.[4,5] The cell block is prepared by processing the small tissue fragments or sediments retrieved from cytological specimens into paraffin blocks.[6] It is widely accepted that the diagnostic accuracy is increased by cell block techniques.[4]

Case Report

A 77 years old female presented to our OPD with history of abdominal pain for a month. Her general condition was fair. Systemic examination was normal. Blood investigation was within normal limits.

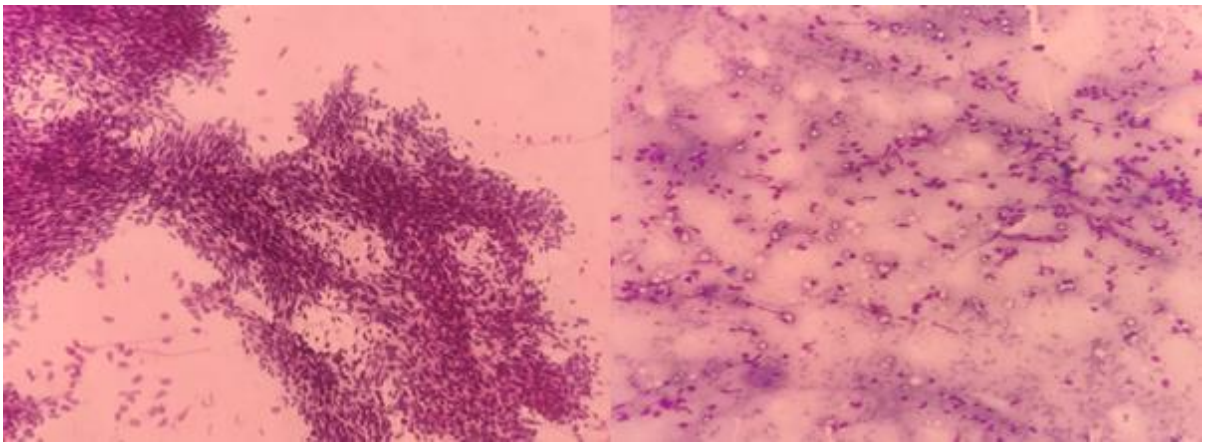

Figure 1: Cytology smears showing spindle shaped tumor cells (Pap stain & Giemsa stain)

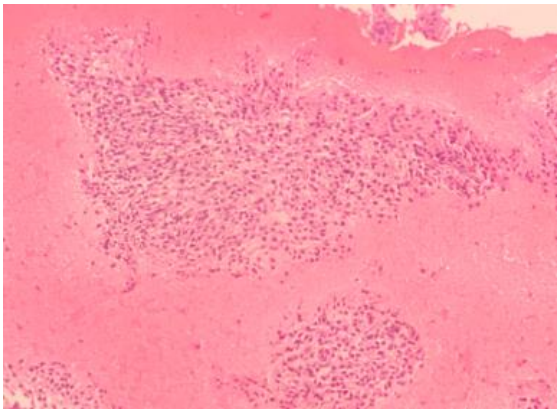

Figure 2: Cell block preparation showing tumor cells arranged in diffuse pattern (H&E stain)

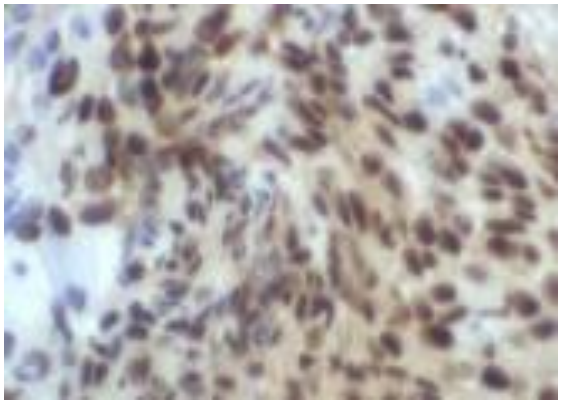

Figure 3: Immunopositivity for TLE1

Blood investigation was within normal limits. There was past history of pulmonary tuberculosis for which she had taken complete medication. CT scan of abdomen and pelvis was done and showed multiple conglomerated necrotic lymph nodes in the retroperitoneum in the para-aortic region measuring 8 x 5 cm suggestive of metastatic lymph nodes. CT guided FNAC was done and the smears were highly cellular comprising spindle shaped cells **(Figure 1)** with plump to elongated nuclei, coarse chromatin and moderate pleomorphism. Mitotic activity was seen with some atypical ones. The cytological features in conjunction with cell block preparation **(Figure 2)** findings were suggestive of malignant spindle cell lesion. The differential diagnoses included retroperitoneal sarcoma and gastro-intestinal stromal tumor.

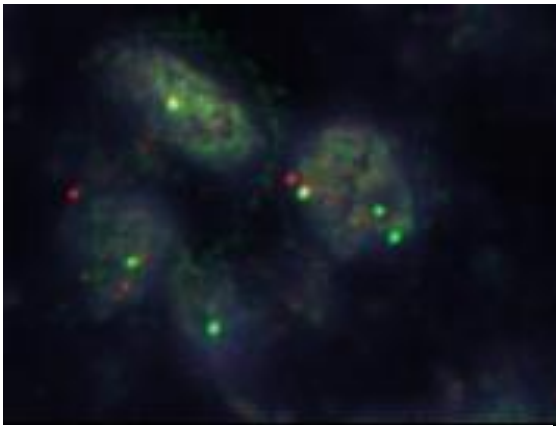

Figure 4: FISH showing SS18 gene rearrangement

Immunohistochemistry was performed on cell block sample which showed positivity for TLE1 (Figure 3), CK and negativity for desmin, SMA, S100, CD117, CD45, FLI1, MDM2, CDK4 and MyoD1; suggestive of synovial sarcoma. The diagnosis of synovial sarcoma was further confirmed by FISH studies showing SS18 gene rearrangement (Figure 4). Detail clinical and radiological evaluation was done but did not reveal any other significant findings. The final diagnosis was given as primary monophasic retroperitoneal synovial sarcoma. The patient was given three cycles of AIM regimen (Adriamycin, Ifosfamide and mesna). Then the patient was lost to follow-up.

Discussion

Synovial sarcoma is a well-defined entity with uncertain histogenesis. Despite its name, there is no evidence that tumor arises from or differentiates towards synovium.[7]

Preoperative diagnosis of retroperitoneal synovial sarcoma is challenging due to its nonspecific imaging features mimicking benign lesions or metastatic nodes.[1,3] However, these tumors appear hypodense on CT scan with poor central enhancement due to necrosis, hemorrhage, cystic change and about 30% cases show intra tumoral calcification.[8] Pathological examination remains the mainstay for the diagnosis of such tumors.

Synovial sarcoma is divided into biphasic, monophasic spindle, monophasic epithelial and poorly differentiated types.[2,3] Monophasic spindle synovial sarcoma is the most predominant subtype which can be confused with other spindle shaped tumors like fibrosarcoma, leiomyosarcoma, MPNST, spindle cell melanoma, GIST and sarcomatoid carcinoma.[9,10]

Cell block increases the diagnostic accuracy owing to the paramount role they play in ancillary tests like immunohistochemistry and molecular studies like FISH.[6] Immunohistochemistry was done in cell block sample and showed positivity for CK, CD99 and TLE; suggestive of SS.[2,10] Negativity for desmin, S100, CD117, SMA and CD45 ruled out other tumors with spindled morphology.[11] However, the golden diagnostic tool in 90% cases is SS18 gene rearrangement which has been demonstrated in our case in cell

block sample highlighting the immense role of cell block in diagnosis.[3,4,5,10]

Conclusion

Even though primary retroperitoneal synovial sarcoma is a rare entity, it should be kept in differential diagnoses of retroperitoneal mass. The diagnosis is based on histopathological, immunohistochemical and molecular studies. Our study has highlighted the significance of cell block in diagnosis of tumors.

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