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A RARE CASE REPORT OF UNDIFFERENTIATED PLEOMORPHIC SARCOMA WITH CUTANEOUS METASTASES DIAGNOSED ON CYTOLOGY.

Pathology

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ABSTRACT

Soft Tissue Sarcomas (STS) are rare tumours with dismal prognosis. Undifferentiated Pleomorphic Sarcoma (UPS) was formerly called Malignant Fibrous Histiocytoma (MFH) and reclassified by the World Health Organization (WHO) in 2013. UPS account for 5% of adult soft tissue sarcomas. It usually occurs in elderly with a male predominance and is seen on extremities. Despite of it being highly metastatic, cutaneous metastasis is found to be very rare. Hence we present this case of soft tissue UPS of thigh metastasizing to lung, other soft tissues as well as cutaneous locations diagnosed by Fine Niddle Aspiration Cytology (FNAC).

KEYWORDS

Undifferentiated Pleomorphic Sarcoma, metastases, rare

INTRODUCTION

Among all the cancers, soft tissue sarcomas are rare accounting for < 1%, of which Undifferentiated Pleomorphic Sarcoma (UPS) being the 4th most common subtype.[1] The former nomenclature of Malignant Fibrous Histiocytoma (MFH) was reclassified by WHO in 2013 as UPS.[2] These tumours are rare with an incidence of about 0.08-1.0 per 1,00,000.[1] UPS has peak incidence between 5th and 7th decade of life.[3] Slight male predominance is seen.[4] The common primary sites are the extremities (60%), trunk (19%), retroperitoneum (15%), and head and neck (9%).[5] The local recurrence rate of UPS ranges between 19% to 31%, and the rate of metastasis is between 31% to 35% even after tumour resection with a clear resection margin. Despite the high distant metastasis rate, cutaneous metastasis is very rare, its rate being 0.6% to 1.5%.[6] As there is lack of definite clinical and radiological signs for early diagnosis, pathological examination plays a key role in diagnosis and management of patient. Due to its low 5 year survival rate, (14%) [7] early diagnostic modality like Fine Needle Aspiration Cytology (FNAC) can be incredibly beneficial. Hence we present this case of soft tissue undifferentiated pleomorphic sarcoma of thigh metastasizing to skin and other site diagnosed by FNAC.

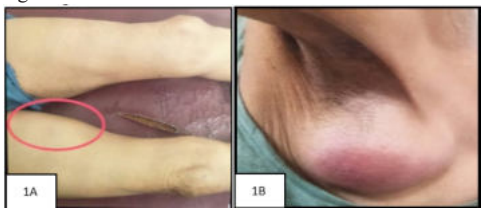
MATERIALS AND METHODS

FNAC procedure was done from multiple sites which included swellings in right thigh, left arm, left axilla, left inguinal region and cutaneous swelling over left forehead. The FNAC sites were cleaned by spirit, and aspirated with 23G needles and 5 cc syringes. Needles were inserted at convenient angles to the swellings and multiple hits were made with sufficient negative pressure followed by removal of needle and application of firm pressure to avoid bleeding and hematoma formation. The aspirate was hemorrhagic with admixed myxoid material which was smeared on glass slides. Few of the slides were wet fixed in 95 % of ethyl alcohol while the rest were air dried. Alcohol fixed smears were stained with Hematoxylin and Eosin (H. & E.) stains and air dried slides were stained with Leishman's and studied under light microscope.

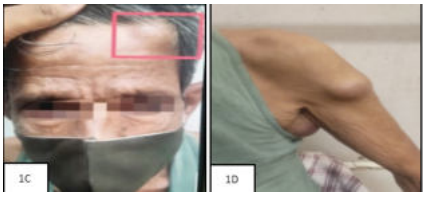
Case Study

Sixty two years, male, a known hypertensive and asthmatic, came with complains of loss of appetite along with loss of weight since 3 months, difficulty in breathing since 2 months, backache since 1 month with axillary and cutaneous swellings noticed since 2 months. He was admitted for the same, underwent radiological investigations and referred to department of pathology with requisition for FNAC of palpable swellings. The chest X-ray revealed a well-defined oval homogenous opacity in the left mid and lower zone in para-cardiac region suggestive of neoplastic etiology. Computed Tomography (CT) scan revealed 6.7x6.7x7.6 cm large heterogeneously enhancing sub-pleural lesion in posterior segment of left lower lobe with multiple necrotic mediastinal lymph nodes and lesions in the left axillary and posterior to right kidney suggestive of primary pulmonary lesion with disseminated metastases. Following this patient underwent Positron Emission Tomography and Computed Tomography (PET-CT) scan which showed high Fluro-deoxy-glucose (FDG) uptake in left lower lobe with scattered nodules in lung, mediastinum, left axilla, abdominal, pelvic, left inguinal lymph nodes, bilateral adrenal, liver, omentum, mesenteric, muscular and skeletal metastases.

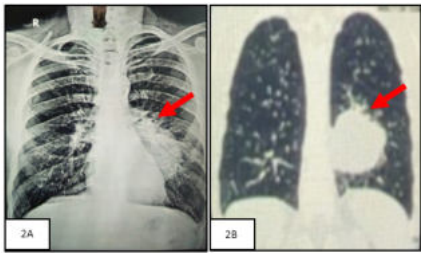
On examination, he had multiple palpable swellings in right thigh, left axilla, left forehead, left arm and left inguinal region. The largest swelling was on medial aspect of upper right thigh, which was diffuse, ill-defined, non-tender, partly mobile, firm and measuring in size of 15x12 cm. Few of the cutaneous swellings were skin fixed. However, on asking the detailed history, he told that thigh lesion was first to be noticed, approximately 6 months back which began as small swelling. Following this, there was appearance of swellings in left inguinal, left axilla, left arm and left forehead in the sequence of occurrence. Aspirate was done from all palpable swellings.The microscopy of all slides showed cellular smears with predominantly singly scattered, non-cohesive oval to round large, pleomorphic cells showing marked nuclear pleomorphism, high nuclear-cytoplasmic ratio (N/C ratio) and vesicular nuclei moderate to marked nucleomegaly. At places, atypical mitosis, bizarre features with tumour giant cells were noted. Few cells with binucleation and cytoplasmic vacuolation were observed. Background revealed amorphous, pink, eosinophilic matrix material admixed with nuclear debris, apoptotic nuclei admixed with blood. There was no evidence of atypical lymphoid cells or lipoblast or rhabdomyoblast or epithelial malignant cells. The above microscopic features were observed in the largest thigh swelling as well as from cutaneous sites. After correlating the clinical, radiological and cytological findings, diagnosis of UPS of right thigh with disseminated including cutaneous metastases was made.



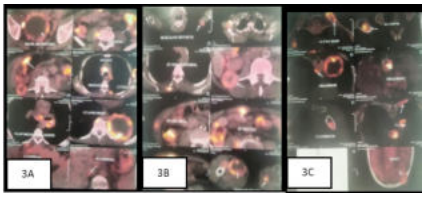
1A- Right thigh swelling – Seen on medial aspect measuring 15x12 cm ,which was diffuse, ill-defined, firm, non-tender and partly mobile.**1B-**Left axillary swelling of size 5x5 cm, well circumscribed, soft to firm, slightly tender, partly mobile, skin fixed with red and inflamed overlying skin.



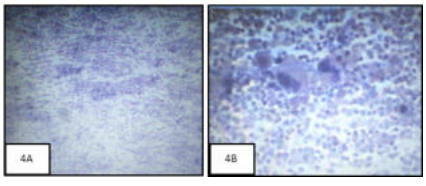
1C- Swelling over left forehead measuring 3x3 cm, well circumscribed, firm, partly tender, immobile, skin fixed.**1D-**Left arm swelling seen on anterior aspect, measuring 3x3 cm, well circumscribed, soft to firm, partly mobile, non skin fixed.



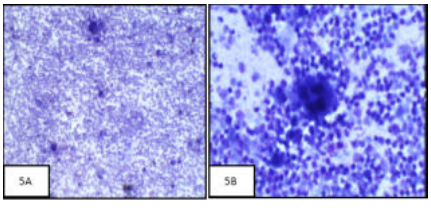
2A- Chest X-ray – A well defined oval homogenous opacity in the left mid and lower zone in para-cardiac region suggestive of neoplastic etiology. **2B-** CT scan revealed 6.7x6.7x7.6 cm large heterogeneously enhancing sub pleural lesion in posterior segment of left lower lobe.



3A,3B,3C- PET-CT scan – High FDG uptake in left lower lobe with scattered nodules in lung, mediastinum, left axillary, abdominal, pelvic, left inguinal lymph nodes, bilateral adrenal, liver, omental, mesenteric, muscular and skeletal metastases.



4A: Singly scattered, non-cohesive oval to round cells with marked nuclear pleomorphism. (H. & E., 10X) **4B:** Pleomorphic cells with high N/C ratio, vesicular nuclei, atypical mitosis, at places bizarre cells. (H. & E., 40X)



5A: Singly scattered, non-cohesive oval to round cells with high N:C ratio and marked nuclear pleomorphism (Leishman's stain, 10X) **5B:** Bizarre cells and tumour giant cells noted.(Leishman's stain, 40X)

DISCUSSION

In the 1960s, MFH was first described by O'Brien and Stout. [8] It is the malignant soft tissue tumour of mesenchymal origin which often metastasizes and recurs 12 to 24 months after diagnosis.[3]

The local recurrence rate of UPS is 19% to 31%, with the metastasis rate is 31% to 35% even after tumour resection with a clear resection margin. Despite the high distant metastasis rate, cutaneous metastasis is very rare. Meanwhile, the cutaneous metastasis rate is 0.6% to 1.5%. Cutaneous metastatic undifferentiated pleomorphic sarcoma carries very poor prognosis hence its diagnosis is of great importance. [6] It has poor prognosis which is not related to the tumour cell abnormality, but to the tumour size, grade, location, pathological type, immunosuppression status and patient's age. [3] The available literature on five and ten year survival rate suggest tumour with high grade, deeper location and size >5cm have the worst overall 10-year survival rates (38%). [9] Also according to Grimer [10], the risk of metastases in sarcoma increases linearly as tumour enlarges. This mandates an early definitive diagnosis to prevent its spread and avoid recurrence. Despite of above known fact, there is unavoidable delay in diagnosis of sarcoma as patients present at later stage due to painless swellings and deeper location.

In case of soft tissue tumours, differentiation of benign from malignant tumours is a pre-requisite. FNAC is rapid, easily available, cheap and low-morbid alternative to open biopsy or needle core biopsy for soft tissue masses. Exact sub-classification is not always necessary for planning the therapy of adult soft tissue sarcomas. Just a reliable diagnosis of sarcoma combined with radiological data and clinical staging is sufficient when treatment includes primary surgery. [11] In this era of modern and advanced diagnostic modalities, FNAC is often sidelined. But detailed clinico-radiological correlation, proper technique of FNAC and accurate morphological evaluation on light microscopy makes FNAC an excellent diagnostic tool in such cases.

CONCLUSIONS

Soft tissue sarcomas are rare, rapidly growing and metastasizing tumours. Hence diagnosis at early stage is necessary for early intervention to avoid untoward complications. In such case scenario, FNAC should be thought of as an initial diagnostic modality which is accurate, rapid, sensitive and inexpensive procedure. Aspirate from primary and metastatic site confirm diagnosis, helping to streamline the early timely management of patient. It helps in confirmation of diagnosis thus helping to streamline the management of patients.

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