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Navigating Undifferentiated Pleomorphic Sarcoma Through Immunohistochemical Analysis

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Abstract: ***Soft tissue sarcomas represent less than 1% of cancers[2], with undifferentiated pleomorphic sarcoma being a rare subtype at 0.08-1 per 100,000 people.[1] This case underscores its rarity and diagnostic difficulty. Undifferentiated Pleomorphic Sarcoma commonly affects men in their 6th and 7th decades, typically in the lower extremities.[3]***

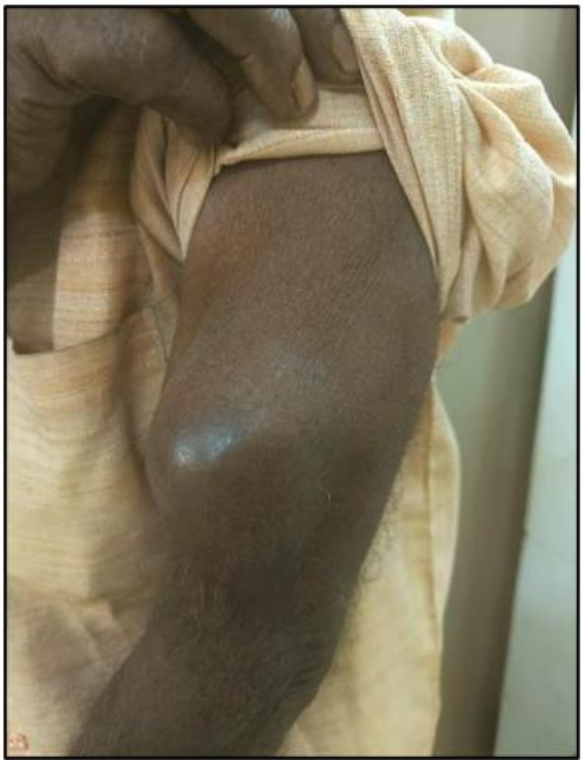
Keywords: Soft tissue sarcoma, Poorly differentiated malignant lesion, Undifferentiated pleomorphic sarcoma, High grade pleomorphic sarcoma, Malignant peripheral nerve sheath tumor.

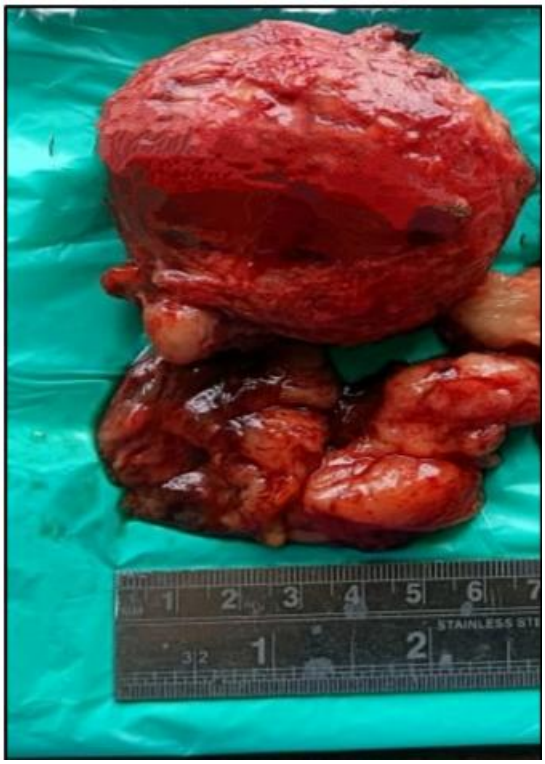
1. Case History

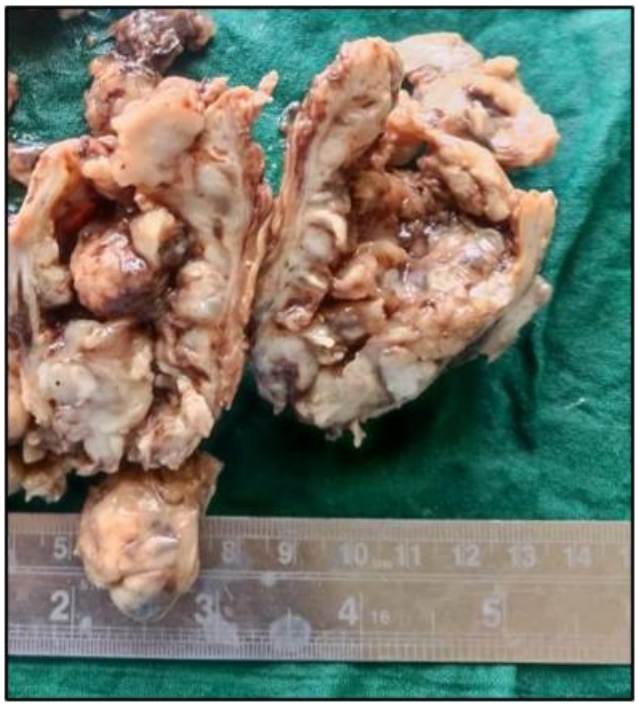
An 88-year-old male presented with rapidly growing 8x5 cm swelling on the left upper arm since four months which is increasing in size, associated with pain and itching. Cytology suggested a poorly differentiated malignant lesion, likely undifferentiated sarcoma.

Gross:
Received grey brown globular soft tissue mass measuring 8x8x3cm.External surface is congested and grey white to grey brown. Cut surface shows grey white to variegated appearance with foci of necrosis.

Gross Pictures:

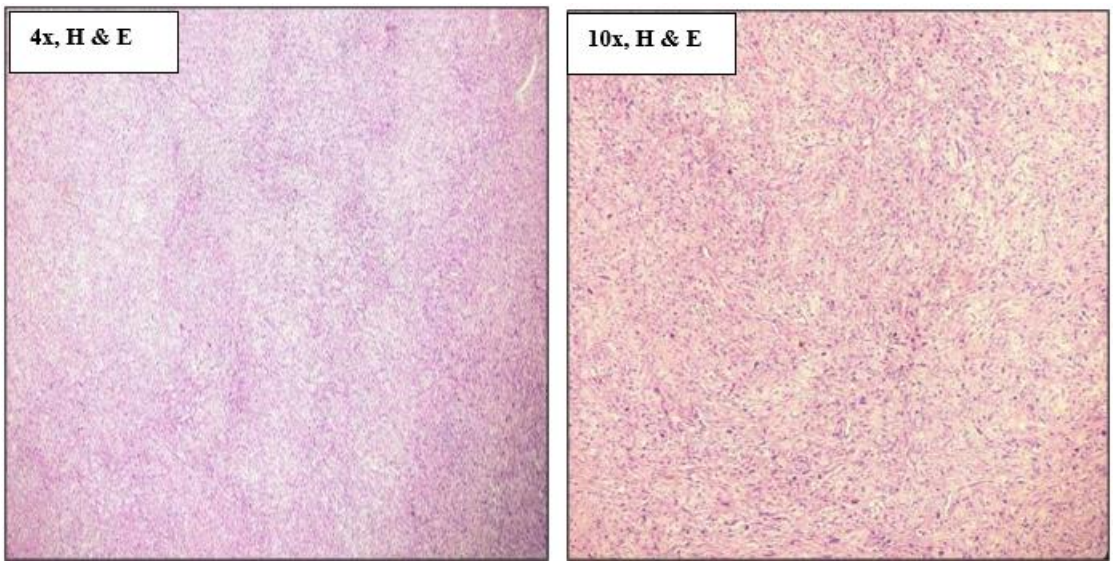


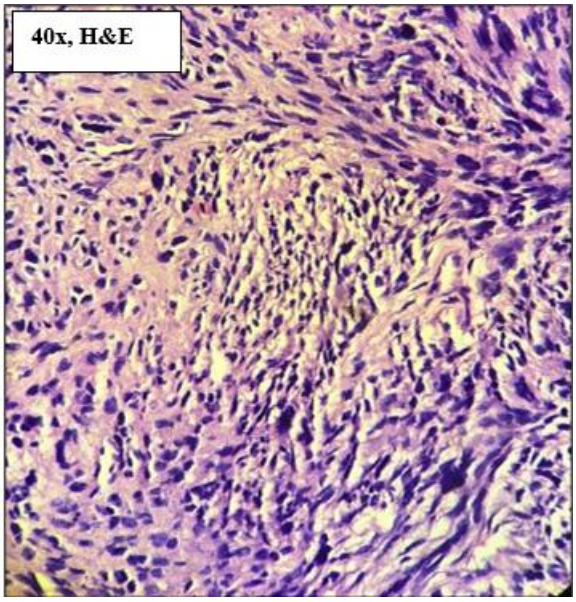




Microscopy:
Tumor cells are arranged in fascicles exhibiting high pleomorphism, oval to spindle-shaped hyperchromatic nuclei showing increased nuclear to cytoplasmic ratio and buckling of nuclei. Additional findings included focal perivascular accentuation, ischemic necrosis, hyper and hypocellular areas, and increased mitotic activity.

Microscopic Pictures:





2. Discussion

High-grade pleomorphic malignant tumors that do not express immunohistochemical markers indicative of a specific line of differentiation are classified as undifferentiated pleomorphic sarcomas (formerly malignant fibrous histiocytomas) [4].

These tumors most frequently arise in individuals in their sixth or seventh decade of life and typically occur in the extremities, retroperitoneum, or head and neck regions.[5] Clinically, patients with undifferentiated high-grade pleomorphic sarcoma (UHPS) often present with a gradually enlarging, non-tender, firm soft-tissue mass.[5]

The main differential diagnoses for undifferentiated pleomorphic sarcoma (UPS) include the various subtypes of soft tissue sarcomas (STS)[2]. These can be effectively excluded through clinical evaluation, comprehensive physical examination, and detailed histopathological assessment, particularly with the use of appropriate immunohistochemical markers.[2]

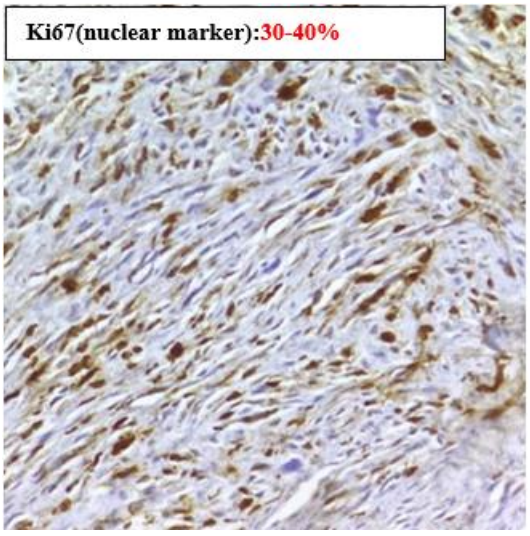
Malignant peripheral nerve sheath tumors (MPNSTs), accounting for approximately 2% of all soft tissue sarcomas, are rare and genomically unstable malignant neoplasms. [6] Malignant peripheral nerve sheath tumors occur most commonly in the trunk and extremities followed by head and neck areas.[7]Microscopy shows spindle shaped cells arranged in fascicles with perivascular accentuation of tumor cells.[7]

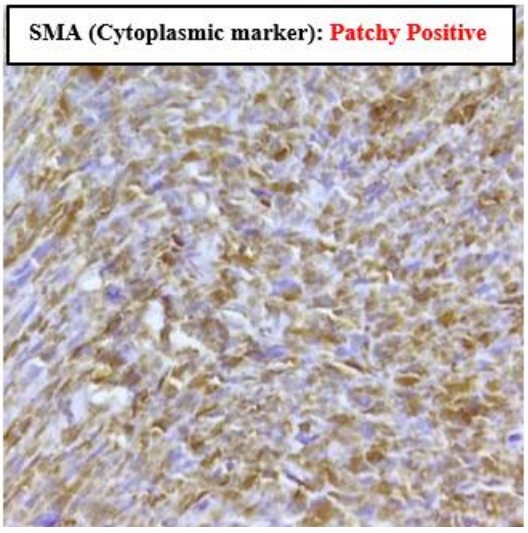
A panel of IHC markers are done to differentiate Undifferentiated pleomorphic sarcoma from other soft tissue tumors.[3]

In our case the following IHC markers are done:
SMA, ki67, S-100, SOX 10, Desmin, H-Caldesmon, Myogenin, CDK 4, MDM 2.

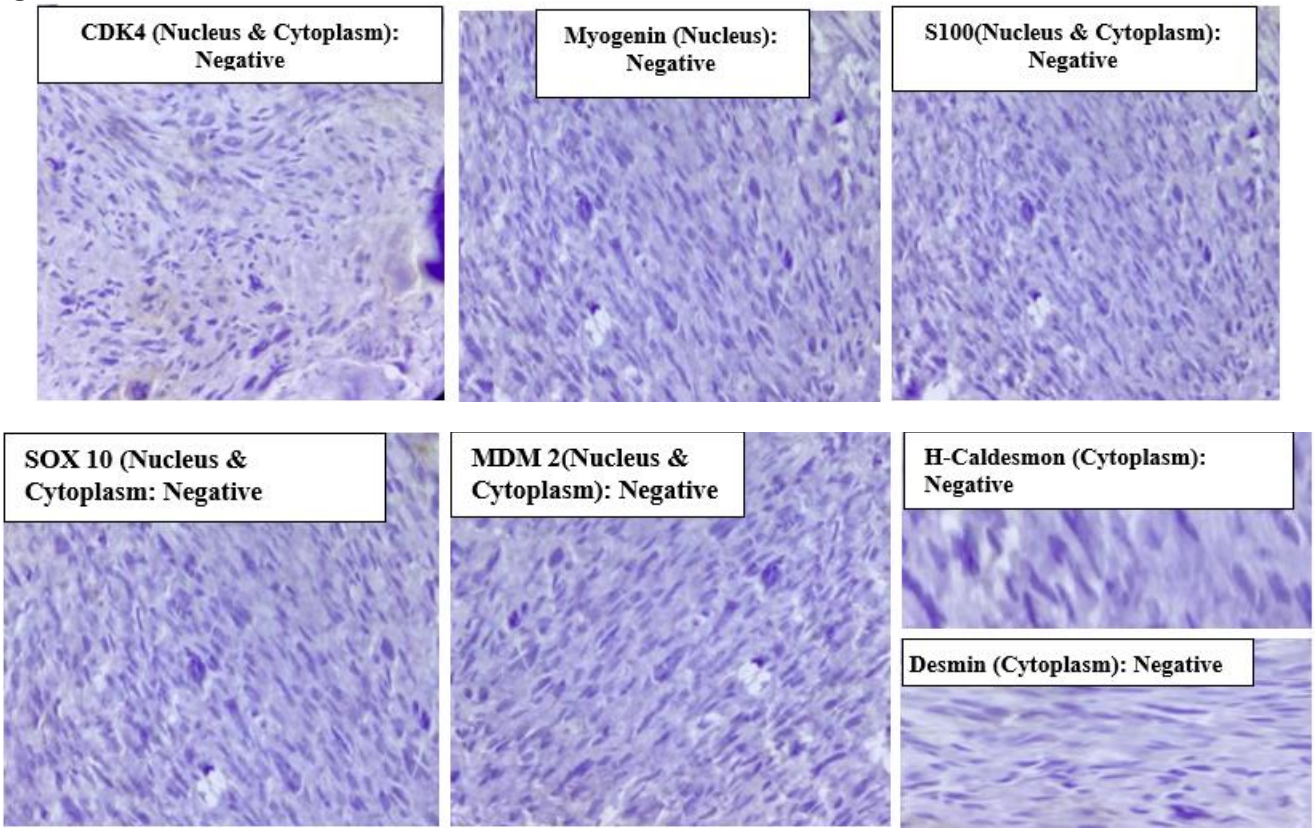
Only SMA showed patchy positivity and ki67 showing 30-40% positivity indicating that it is an aggressive tumor .In our case S100, SOX10 are negative excluding nerve sheath tumors. Desmin and H-Caldesmon are negative excluding smooth muscle tumors. CDK4 and MDM2 are negative excluding well differentiated liposarcoma.

Positive Markers





Negative Markers



Differential Diagnosis

1) Malignant peripheral nerve sheath tumor.
2) Undifferentiated pleomorphic sarcoma.

3. Conclusion

Undifferentiated Pleomorphic Sarcoma is a diagnosis of exclusion. Therefore, IHC is necessary to exclude other lineages before coming to the diagnosis of Undifferentiated Pleomorphic Sarcoma.

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