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EDITORIAL

The Sinewy Vortex-Undifferentiated Pleomorphic Sarcoma

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Undifferentiated pleomorphic sarcoma is a commonly discerned soft tissue sarcoma, previously designated as malignant fibrous histiocytoma with pleomorphic, myxoid or giant cell subtypes. Undifferentiated pleomorphic sarcoma is comprised of fibroblasts, myofibroblasts and histiocyte-like cells. Tumefaction may emerge adjacent to orthopaedic implants or following irradiation.

Preponderantly a diagnosis of exclusion, copious quantities of tissue require sampling in order to eliminate dedifferentiated neoplasms or differentiation into pertinent mesenchymal components.[1,2]

Undifferentiated pleomorphic sarcoma may represent as a terminal stage of diverse sarcomas manifesting unifying morphological features of cellular and nuclear pleomorphism and a storiform tumour configuration.[1,2]

Undifferentiated pleomorphic sarcoma commonly emerges within adults > 50 years. A slight male preponderance is observed.[1,2]

The enlarged, deep-seated, progressive neoplasm is commonly confined to lower extremities whereas sites such as retroperitoneum, head and neck or breast are infrequently incriminated.[1,2]

Neoplastic cells of undifferentiated pleomorphic sarcoma demonstrate a complex triploid or tetraploid karyotype. Upon gene expression profiling, tumefaction resembles diverse sarcomas. Genomic alterations within G1/S checkpoint genes are frequently discerned.[1,2]

Upon gross examination, tumefaction appears as an enlarged, lobulated, soft tissue mass which may appear circumscribed. Foci of calcification or ossification may be observed. Subcutaneous lesions demonstrate a magnitude of ≤ 5 centimetres whereas retroperitoneal neoplasms may extend to ~ 20 centimetres.[1,2]

Cut surface is fleshy and fibrotic with focal areas of haemorrhage, necrosis or myxoid alterations.[1,2]

Cytological examination demonstrates singular cells admixed with enlarged tissue fragments depicting a storiform pattern. Tumor cells appear as spindle-shaped, plasmacytoid or pleomorphic with malignant nuclear morphology and are admixed with numerous multinucleated tumor giant cells.[1,2]

Upon microscopic examination, the variably cellular neoplasm exhibits irregular fascicles and a distinctive storiform pattern wherein cells appear to emanate from a singular, centric focus.[1,2]

Tumour cells appear pleomorphic, bizarre and display significant cellular atypia with prominent variation in cellular and nuclear magnitude.

Neoplastic cells appear imbued with foamy cytoplasm. Surrounding stroma is predominantly collagenous and appears invaded with chronic inflammatory cells. Innumerable mitotic figures are intermingled with atypical mitosis.[1,2]

Tumour parenchyma may be infiltrated by multinucleated giant cells wherein a preponderance of giant cells incurs a terminology of giant cell malignant fibrous histiocytoma. Foci of metaplastic bone or cartilage may be discerned.[1,2]

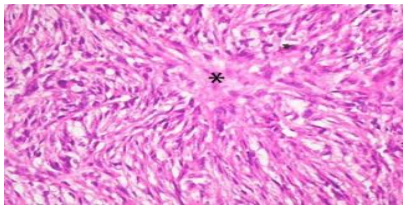

Figure 1: Undifferentiated pleomorphic sarcoma depicting a storiform pattern of pleomorphic fibroblastic and my fibroblastic cells imbued with eosinophilic cytoplasm, nuclear atypia and mitotic figures.[5]

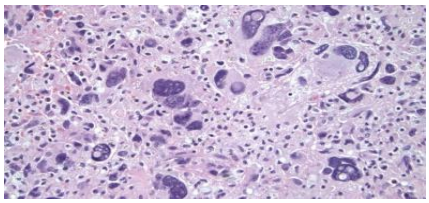

Figure 2: Undifferentiated pleomorphic sarcoma depicting pleomorphic fibroblasts and myofibroblasts intermingled with numerous multinucleated giant cells and foamy macrophages.[6]

Neoplastic cells are devoid of specific cellular lineage and tumefaction is comprised preponderantly of undifferentiated fibroblastic, my fibroblastic or primitive mesenchymal cells. Few tumour cells may depict phagocytic properties.[3,4]

Table: Histologic Grade of Soft Tissue Sarcoma.[2,3]

Tumour Differentiation	**Description**
1	Simulating normal adult mesenchymal tissue
2	Certain histological subtypes
3	Embryonal, Ewing's, synovial, primitive neuroectodermal or undifferentiated sarcoma
Mitotic Activity	
1	0-9 mitosis/10 high power fields
2	10-19 mitosis/10 high power fields
3	≥20 mitosis/10 high power fields
Tumour Necrosis	
0	Absence of necrosis
1	<50% necrosis
2	≥50% necrosis

Undifferentiated pleomorphic sarcoma is immune reactive to vimentin, alpha-1-antitrypsin, alpha-1-antichymotrypsin, factor XIIIa, CD68, CD10, CD34, CD99 along with an exceptional, aberrant reactivity to Melan A.[3,4]

Tumour cells are immune non-reactive to keratin, melanocytic immune markers as CD45 or S100 protein and immune markers of myogenic differentiation.[3,4]

Undifferentiated pleomorphic sarcoma requires segregation from neoplasms such as anaplastic large cell lymphoma, atypical fibroxanthoma, pleomorphic leiomyosarcoma, histiocytoid leprosy, pleomorphic liposarcoma, giant cell malignant fibrous histiocytoma, inflammatory malignant fibrous histiocytoma, myxofibrosarcoma, metastatic renal cell carcinoma, pleomorphic rhabdomyosarcoma, angiosarcoma, fibrosarcoma, dermatofibrosarcoma protuberans, osteosarcoma or malignant peripheral nerve sheath tumor.

Also, tumor metastases emerging from diverse primary neoplasms, desmoplastic melanoma or spindle-cell squamous cell carcinoma may simulate undifferentiated pleomorphic sarcoma.[3,4]

Segregation of cutaneous undifferentiated pleomorphic sarcoma from atypical fibroxanthoma or dermatofibrosarcoma protuberans may be challenging.[3,4]

Computerized tomography exhibits nodular and peripheral enhancement of solid tumour segments. Minimally attenuated centric zones may correspond to foci of preceding haemorrhage, necrosis or myxoid alterations. Attenuation of adipose tissue fragments is absent. Heterotopic bone may be configured.[3,4]

Undifferentiated pleomorphic sarcoma arising within extremities can be appropriately alleviated with surgical extermination and removal of broad perimeter of uninvolved soft tissue. Adjuvant or neoadjuvant radiation therapy and chemotherapy is frequently employed.[3,4]

Borderline neoplasms depicting microscopic features intermediate to benign fibrous histiocytoma and undifferentiated pleomorphic sarcoma can be optimally subjected to localized surgical eradication with excision of wide margin of uninvolved tissue.[3,4]

Localized tumour reoccurrence is observed. Distant metastasis may occur within pulmonary parenchyma or regional lymph nodes.

Undifferentiated pleomorphic sarcoma exhibits a 5-year proportionate survival of ~60% wherein survival is variable and superior in diverse subtypes as dedifferentiated liposarcoma.

Superior prognostic outcomes are obtained with therapeutic intervention of miniature, superficial, low-grade neoplasms.[3,4]

Conclusion

Undifferentiated pleomorphic sarcoma is a common soft tissue sarcoma comprised of fibroblasts, myofibroblasts and histiocyte-like cells, previously designated as malignant fibrous histiocytoma with pleomorphic, myxoid or giant cell subtypes. The enlarged, deep-seated, progressive neoplasm is commonly confined to lower extremities whereas sites such as retroperitoneum, head and neck or breast are infrequently incriminated. The variably cellular neoplasm exhibits irregular fascicles and a distinctive storiform pattern wherein cells appear to emanate from a singular, centric focus. Undifferentiated pleomorphic sarcoma is immune reactive to vimentin, alpha-1-antitrypsin, alpha-1-antichymotrypsin, factor XIIIa, CD68, CD10, CD34, CD99 along with an exceptional, aberrant reactivity to Melan A. Undifferentiated pleomorphic sarcoma requires segregation from neoplasms such as anaplastic large cell lymphoma, atypical fibroxanthoma, pleomorphic leiomyosarcoma, histiocytoid leprosy, pleomorphic liposarcoma, giant cell malignant fibrous histiocytoma, inflammatory malignant fibrous histiocytoma, myxofibrosarcoma, metastatic renal cell carcinoma, pleomorphic rhabdomyosarcoma, angiosarcoma, fibrosarcoma, dermatofibrosarcoma protuberans, osteosarcoma or malignant peripheral nerve sheath tumor.

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Conflict of Interest

Author declares that there is no conflict of interest.

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5. Image 1 Courtesy: Science direct.
6. Image 2 Courtesy: My pathology report.com