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RESEARCH ARTICLE

A RECURRING CASE OF MALIGNANT FIBROUS HISTIOCYTOMA (UNDIFFERENTIATED PLEOMORPHIC SARCOMA) IN THORACO-LUMBAR REGION

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ABSTRACT

Malignant Fibrous Histiocytoma (UNDIFFERENTIATED PLEOMORPHIC SARCOMA)-storiform pleomorphic type is the classic type, consists of plump spindle shaped cells arranged in short fascicles in a cartwheel or storiform pattern. Histopathological along with IHC is the main way for diagnosis. Treatment is surgical resection along with radiotherapy and chemotherapy **Presentation:** We present an unexpected diagnosis of malignant fibrous histiocytoma in a recurrent swelling of subcutaneous plane (thoraco-lumbarregion)of a specimen excised from a 32 year old female. **Impression:** Malignant Fibrous Histiocytoma-storiform pattern.

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INTRODUCTION

Malignant fibrous histiocytoma is a soft tissue sarcoma with fibroblastic and histiocytic differentiation occurring in the late adult life between the age 50 and 70 years. It is common in extremities, especially thighs. The classical form of UPS has both storiform and pleomorphic features, with plump spindle shaped cells with marked cytologic atypia arranged in tight whorls along withmitotic figures including atypical forms are typically seen. The tumour is prone to local recurrence and has the capacity to metastasize to distant sites [3] The clinicopathologic findings in 200 cases of malignant fibrous histiocytoma (MFH) with follow-up information are presented. Malignant fibrous histiocytoma (UPS)occurred principally as a mass on an extremity (lower extremity 49%, upper extremity 19%) or in the abdominal cavity or retroperitoneum (16%) of adults (peak incidence 61-70 years of age). [4] This paper reports a case of fibrous histiocytoma(undifferentiated sarcoma) in a 32 year old female who had a recurring mass in thoraco-lumbar region.

CASE REPORT

A 32 year old female presented with complaints of recurrent swelling over the thoraco-lumbar region for past 12 years which rapidly increased in size between 2 years. The patient had similar complaints in the past, twice for which surgical resection was done. Physical examination showed an emaciated female with a massive swelling over the thoraco-lumbar region of size 15*20*3cm.

External surface: Multilobated, irregular surface, irregular margins and edges, no visible pulsation and no impulse on coughing, no complaints of discharge from the swelling, no warmth and no tenderness. Variable in consistency, plane of swelling is subcutaneous.

USG ABDOMEN: Multiple well defined necrotic soft tissue lesion located in subcutaneous plane, internal septation present, increased vascularity present.

MRI DL SPINE: Mesenchymal soft tissue tumour. Wide local excision done and a variable mass was send for histopathological examination.



Figure 1. Gross picture

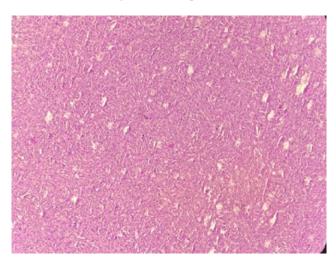


Figure 2. Hematoxylin and eosin (HE) section at 10x magnification

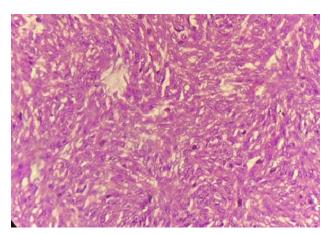


Figure 3. Hematoxylin and eosin (HE) section at 40xmagnification

HISTOPATHOLOGICAL FINDINGS: A cellular tumour composed of interlacing bundles of spindle cells arranged in a storiform pattern. In between histiocytes, inflammatory cells are also seen. Spindle shaped cells arranged in stori form pattern. Increased mitotic activity with few abnormal mitosis seen. The final diagnosis of storiform pattern of malignant fibrous histiocytoma of the thoracolumbar region was made.

DISCUSSION

- The undifferentiated pleomorphic sarcoma (UPS) —
 previously known as malignant fibrous histiocytoma, is a
 high-grade aggressive soft-tissue sarcoma (STS).
 Mesenchymal stem cells are the most probable origin of the
 tumor, instead of histiocytes as previously thought^[1]
- It affects soft tissues, bones, retroperitoneum, and metastasize to several organs. The previous reports suggested it was the most common soft-tissue sarcoma (STS) in the adult population^{-[2]}
- Previous subclassifications of MFH into storiform, pleomorphic, myxoid, giant cell, and angiomatoid variants^[2]
- On Gross examination, UPS is often white to pale yellow in colour, with central hemorrhage and necrosis ^[5].
- It is characterized histologically by high cellularity, marked nuclear pleomorphism, increased mitotic activity (atypical mitoses), along with areas containing spindle cell morphology ^[5]. Necrosis is the common feature of high-grade lesions ^[5].
- In trunk and extremities UPS is staged according to the TNM and histologic grade (G) $^{[6]}$
- Histological grade [G]has the differentiation, mitotic count, and necrosis extension of the tumor, this is stated by the French Federation of Cancer Centers Sarcoma Group (FNCLCC)
- After TNM and G evaluation, the disease is further classified into stages I-IV for therapeutic purposes.

Table 1.

T: Primary Tumor			
Tx	Primary tumor cannot be assessed		
T0	No evidence of primary tumor		
T1	Tumor ≤ 5 cm in highest dimension		
T2	Tumor > 10 cm and ≤ 15 cm		
T3	Tumor > 15 cm in greatest dimension		
N: Regional Lymph Nodes			
N0	Nil regional lymph node metastasis or		
	unidentified lymph node status		
N1	Regional lymph node metastasis		
M: Distant metastasis			
M0	No distant metastasis		
M1	Distant metastasis		
G: Histologic grade			
GX	Grade cannot be assessed		
G1	Total differentiation, mitotic count,		
	necrosis score 2 or 3		
G2	Total differentiation, mitotic count,		
	necrosis score 4 or 5		

Table 2.

Histologic grade	The totality of differentiation, mitotic activity, and		
	extent of necrosis scores.		
Tumor			
differentiation			
1	Closely resembling normal adult mesenchymal		
	tissue.		
2	Certain histologic typing		
3	Embryonal sarcoma, synovial sarcoma, Ewing		
	sarcoma, primitive neuroectodermal, and		
	undifferentiated sarcoma		
Mitotic activity			
1	0-9 mitoses per 10 high power field		
2	10-19 mitoses per 10 high power field		
3	≥ 20 mitoses per 10 high power field		
Tumor necrosis			
0	No necrosis		
1	< 50% necrosis		
2	≥ 50% necrosis		

Table 3.

Stage	T	N	M	G
IA	T1	N0	M0	G1,GX
IB	T2	N0	M0	G1,GX
	T3	N0	M0	G1,GX
	T4	N0	M0	G1,GX
II	T1	N0	M0	G2,G3
IIIA	T2	N0	M0	G2,G3
IIIB	T3	N0	M0	G2,G3
	T4	N0	M0	G2,G3
	Any T	N1	M0	Any G
IV	Any T	Any N	M1	Any G

CONCLUSION

Malignant Fibrous Histiocytoma (Undifferentiated Pleomorphic Sarcoma) has the site predilection of extremities in which thigh region is more common and has a male predisposition. But in my case study the tumour is recurrent in a young female in thoraco-lumbar region even after when excision had been done twice.

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