DEDIFFERENTIATED LEIOMYOSARCOMA UTERUS WITH ANGIOSARCOMATOUS DIFFERENTIATION PRESENTING WITH LUNG METASTASIS

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INTRODUCTION

Dedifferentiated sarcoma is defined as the occurrence of a high grade or undifferentiated tumor from a low-grade/borderline neoplasm with well-defined demarcation between them. It is a rare phenomenon of soft tissue sarcomas and is extremely rare among uterine sarcomas.
Uterine dedifferentiated leiomyosarcoma with angiosarcomatous differentiation is extremely rare. The authors describe herein what is believed to be the first reported case of a dedifferentiated leiomyosarcoma of the uterus with angiosarcomatous differentiation.

CASE HISTORY

A 53-year-old female presented with hemoptysis and dyspnoea. CT chest revealed a left upper lobe mass lesion (m) 2.4 x1.6x3.8 cm .
PET CT showed FDG avid lesions in left upper lobe (Fig. 1) with FDG avid left hilar nodes (Fig. 2).

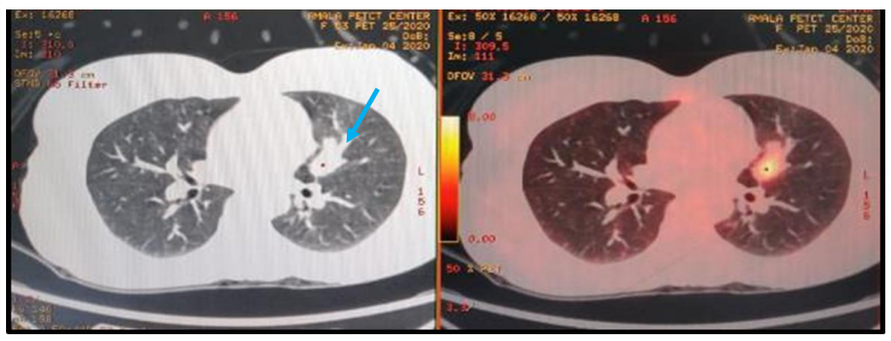

Fig. 1: FDG avid left upper lobe lesion (blue arrow)

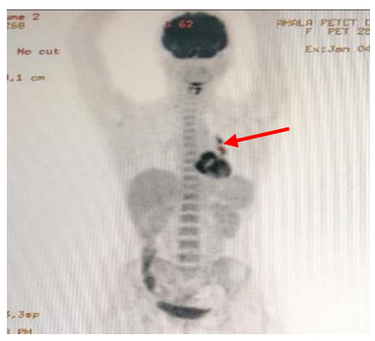

Fig. 2: FDG avid left hilar nodes (red arrow)

She has a history of leiomyosarcoma uterus for which hysterectomy was done 3 months back. So with the provisional clinical diagnosis of metastasis from leiomyosarcoma uterus, left upper lobe of lung resection with lymph node sampling was done.
Gross examination of the lobectomy specimen revealed 3 grey white firm nodular lesions, each measuring 0.8 cm in greatest dimension.

MICROSCOPY

Histopathological examination showed a poorly differentiated neoplasm with an epithelioid morphology (Fig 3&4). Therefore a panel of Immunohistochemical markers were done. The tumour cells were positive for pan-CK (Fig. 5)and negative for Desmin and Melan A in the first panel. The second panel consisted of CK7, CK20, TTF1, Napsin A, Synaptophysin, Chromogranin A, p40, p63. All were found to be negative. In the third panel we put CD31(Fig. 6) and CD34(Fig. 7). The tumour cells were diffusely strongly positive for CD31 and scattered cells only positive for CD34. Hence a diagnosis of epitheliod angiosarcoma was made.

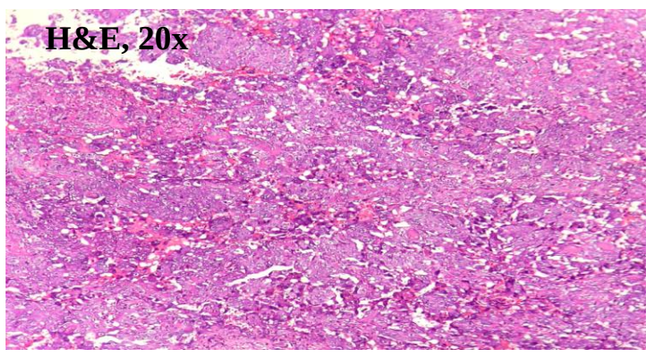

Fig. 3: Poorly differentiated neoplasm in lung

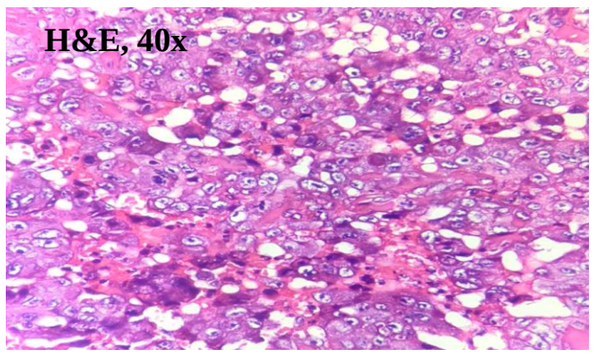

Fig. 4: Tumour cells showing epithelioid morphology.

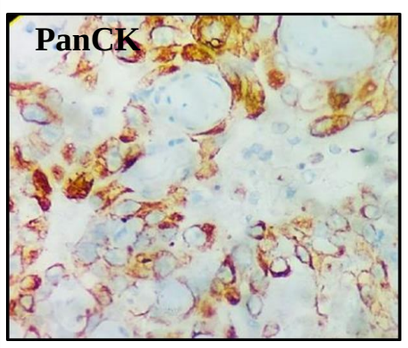

Fig.5, 40x

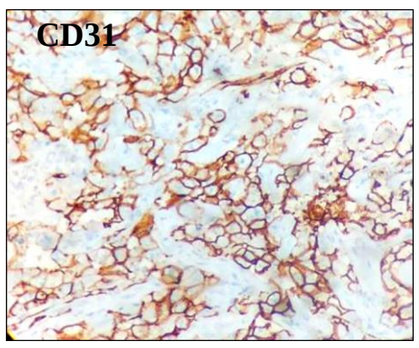

Fig. 6, 40x

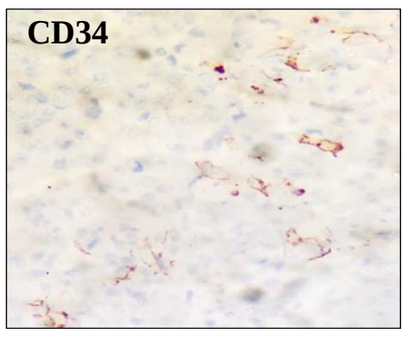

Fig. 7, 40x

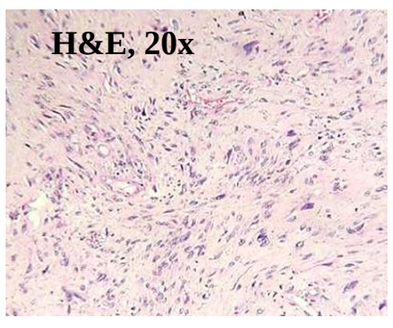

Fig. 8 Leiomyosarcoma

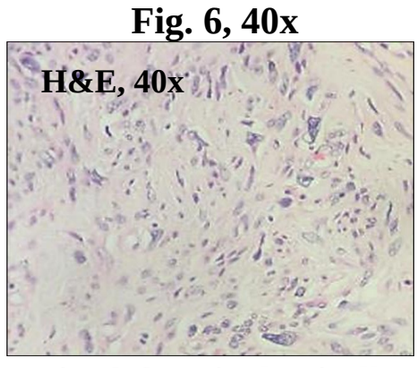

Fig. 9 Atypical spindle cells in LMS

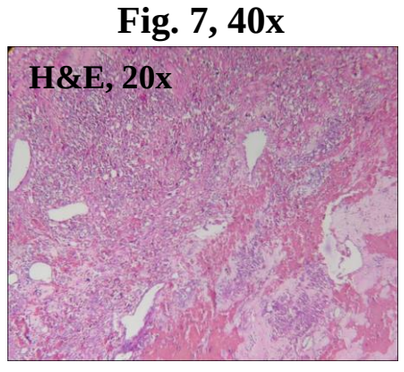

Fig.10 Poorly differentiated areas

The original slides of uterine leiomyosarcoma (LMS) were reviewed. It showed in addition to the conventional leiomyosarcoma (Fig. 8&9), poorly differentiated areas with epithelioid morphology (Fig. 10&11) which earlier was considered to be high grade areas of leiomyosarcoma. On immunohistochemical analysis, these areas turned out to be a neoplasm showing angiosarcomatous differentiation (Table 1) (Fig. 12-14).

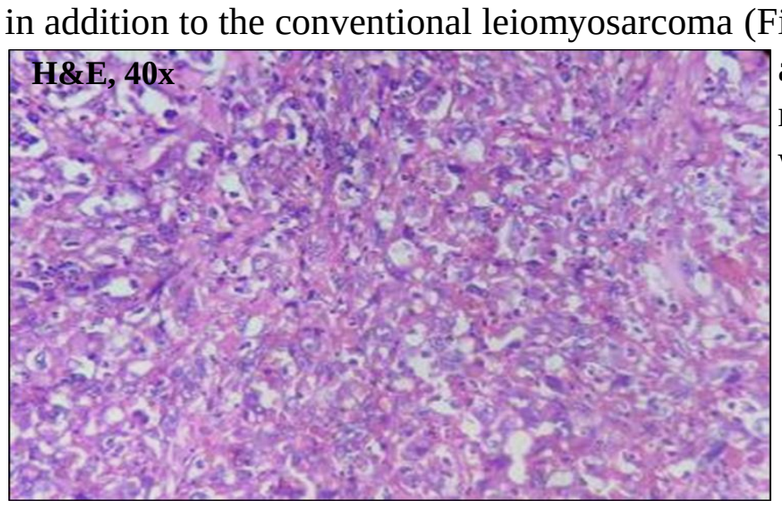

Fig.11 Poorly differentiated areas showing epithelioid morphology

	Desmin, SMA, h-caldesmon	CD31, CD34
Leiomyosarcomatous area	Positive	Negative
Poorly differentiated areas	Negative	Postive

Table 1. IHC analysis of original uterine leiomyosarcoma slides

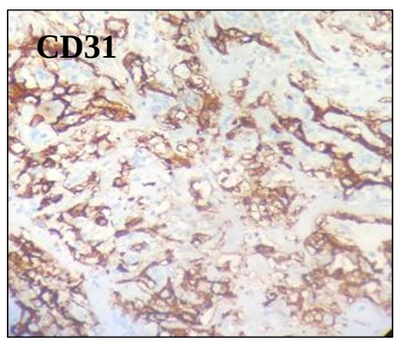

Fig. 12: 40x

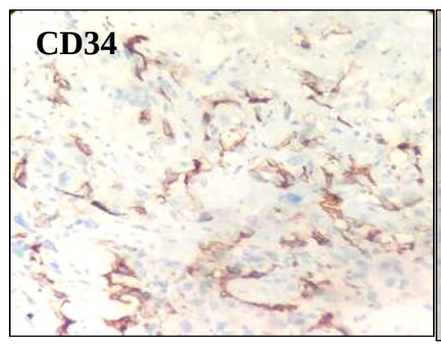

Fig.13: 40x

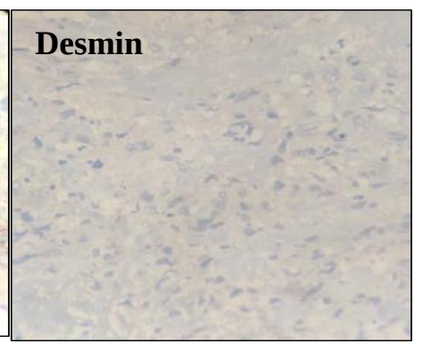

Fig. 14: 40X

DISCUSSION

Leiomyosarcomas are the major soft tissue sarcomas in the pelvis/abdomen, retroperitoneum, trunk and the extremities. Even though the most common sarcoma of the uterus, it accounts for only 1-2% of all malignancies according to the World Health Organization (WHO) classification of Female genital system [1].
Dedifferentiated neoplasms exhibit areas showing high grade histology as well as loss of lineage specific markers in IHC. Dedifferentiation in leiomyosarcoma is very rare and pursue an aggressive clinical course.
To the best of our knowledge, only seven case reports of uterine leiomyosarcoma with dedifferentiation has been reported so far [2-7]. The most common being the one with osteoid heterologous elements. Cases with angiosarcomatous differentiation has not yet been reported.
Uterine dedifferentiated leiomyosarcoma with angiosarcomatous differentiation is extremely rare and can easily be missed. Therefore extensive sampling and use of a panel of immunohistochemical markers are advocated to identify high grade/undifferentiated areas in a well differentiated neoplasm.

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