Case Report

Peritoneal De-differentiated Liposarcoma: A Rare Case Report

Gauri Vinod Hardas*, Lalit Lad, Harshwardhan Bora, Saurabh Prasad

Pathology, KIMS-Kingsway Hospitals, Nagpur, Maharashtra

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***Corresponding Author**:
Dr Gauri Vinod Hardas
gaurihardas05@gmail.com

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Abstract

Peritoneal de-differentiated liposarcoma is a rare malignancy with a poor prognosis. Our patient, 67 years/Male presented with complaints of constipation & distension of abdomen for one month. Initially based on histology, diagnosis of Gastrointestinal Stromal Tumor (GIST) was thought. On PET scan, he had multiple peritoneal deposits & serosal deposits along recto-sigmoid colon and ileal loop which on immunohistochemistry (IHC) proved to be de-differentiated liposarcoma. The patient was given chemotherapy. Patient responded well to treatment. Such peritoneal tumors pose a challenge to the pathologist as only morphology is not of much help & IHC becomes necessity.

Keywords:

De-differentiated liposarcoma, Immunohistochemistry

Introduction

Peritoneum is the rare site for sarcoma. Commonest sarcomas of peritoneum are Gastro Intestinal Stromal Tumor (GIST) and leiomyosarcoma. Liposarcoma of peritoneum is very uncommon. Liposarcoma is the sarcoma originating from adipose tissue. It is classified into different histological types such as well differentiated liposarcoma, myxoid liposarcoma & pleomorphic liposarcoma. De-differentiated liposarcoma is a variant of well differentiated liposarcoma having both lipogenic and non lipogenic components, making its diagnosis really challenging on both radiology and histology.

Case Report

A 67 years old male presented with complaints of constipation and abdominal distension since one month. On examination, patient had moderate ascites for which tapping was done and sample was sent for cytology on three consecutive days which turn out to

be negative for malignancy. His laboratory investigations were normal except raised CA125 levels - 232.8U/ml (reference range 0-35U/ml).

Later, his CECT abdomen was done which showed an ill-defined pelvic mass measuring 5.6x9x8cm, arising from sigmoid colon. There was diffuse omental nodularity & fat stranding causing omental caking. Few soft tissue deposits were noted in the pelvis along right iliac vessels. Further evaluation with PET scan was done. It showed FDG avid peritoneal deposits along serosal surface of recto-sigmoid colon & ileal loops with SUV value 17.9 (Fig-1). Moderate to gross ascites was seen. Hypermetabolic omental soft tissue stranding & multiple peritoneal deposits were noted. Hence based on laboratory & radiological findings clinician initially thought of primary peritoneal carcinoma. Biopsy was taken from one of the peritoneal deposits. On histology, it showed spindle cell tumor arranged in interlacing fascicles (Fig-2). Tumor cells showed moderate nuclear atypia & eosinophilic cytoplasm (Fig-3). Very occasional vacuolated lipoblast like cells seen. Mitotic figures were 5-6/10hpf. Focal area of necrosis seen.

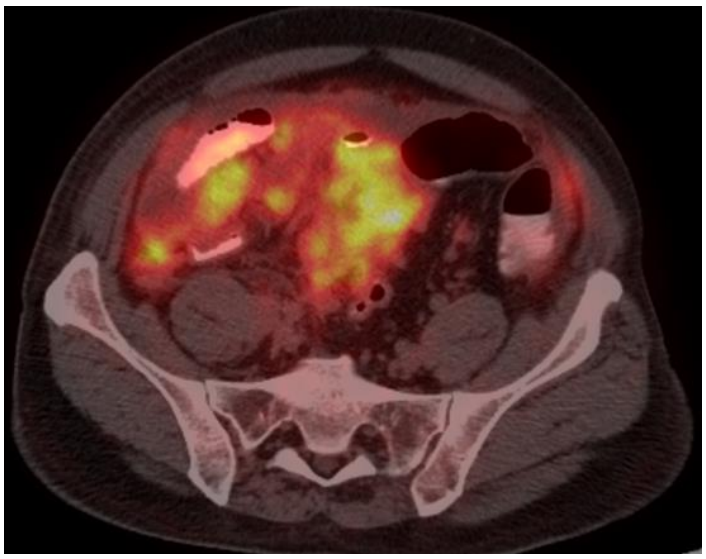

Figure 1 showing FDG avid peritoneal deposits along serosal surface of recto-sigmoid colon & ileal loops with SUV value 17.9

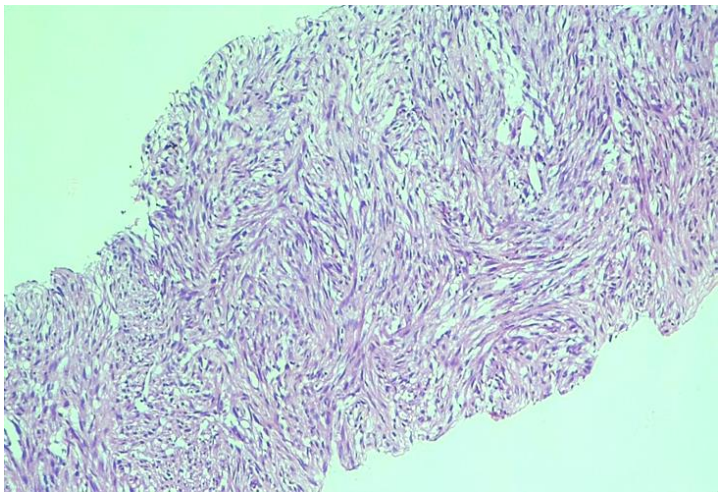

Figure 2 Low power showing malignant spindle cell tumor arranged in interlacing fascicles.

So based on histology, diagnosis of GIST was made. Extensive immunohistochemistry panel was used. Tumor cells were strongly positive for MDM2 (Fig-4), CDK4, focally positive for SMA. Ki67 was 30% (Fig-5). Tumor cells were negative for desmin, S100, CD117, DOG1, h-Caldesmon, ALK, Beta catenin, MSA, SS18 and retained SDH B. Based on histology & immunohistochemistry findings, diagnosis of de-differentiated liposarcoma was made.

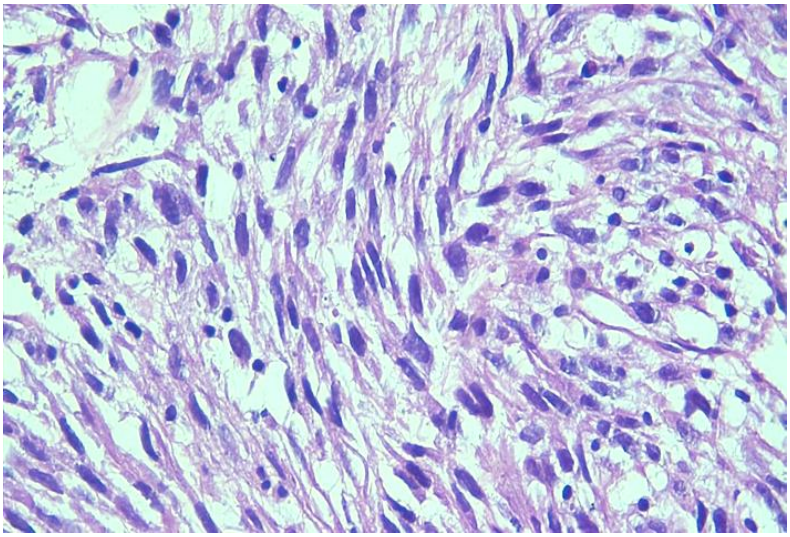

Figure 3 High power showing tumor cells with moderate nuclear atypia & eosinophilic cytoplasm. Rare cytoplasmic vacuolated cells seen

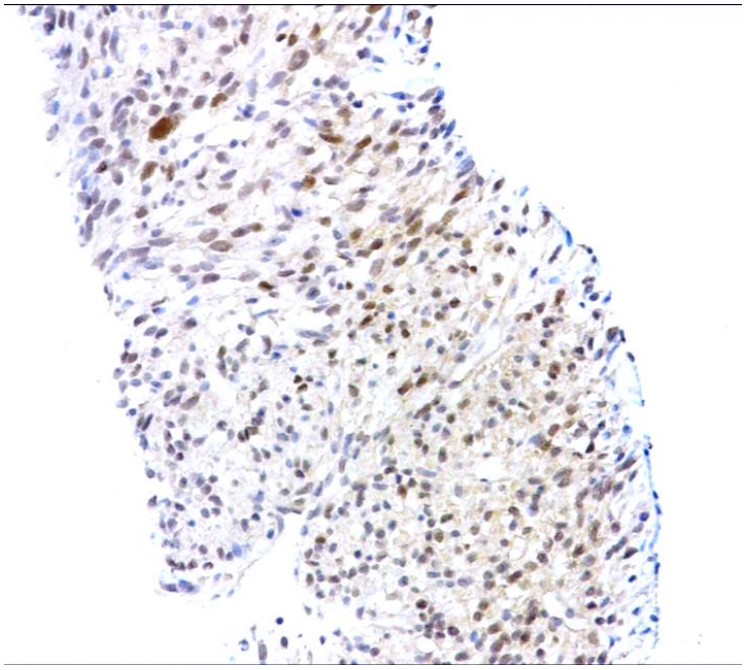

Figure 4 On IHC MDM2- strongly positive

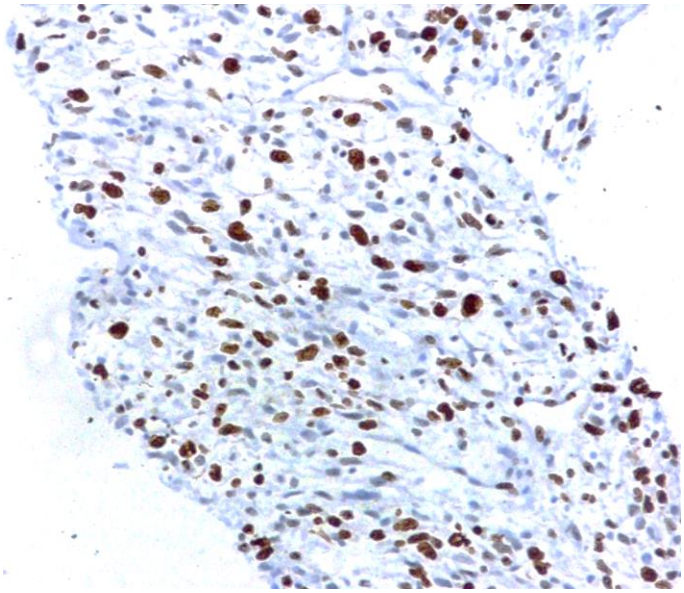

Figure 5 On IHC, Ki67- 30%

Discussion

Liposarcoma is a mesenchymal tumor originating from adipose tissue & is one of the common soft tissue sarcoma [1]. However liposarcoma originating in peritoneal cavity is very uncommon. Liposarcoma is classified into well differentiated, myxoid & pleomorphic type. De-differentiated liposarcoma is considered to be the aggressive form of well differentiated liposarcoma. It has poor prognosis & has more chances of recurrences and can show distant metastasis unlike the well differentiated liposarcoma.[2]

On histology, non lipogenic that is the de-differentiated component resembles fibrosarcoma or undifferentiated pleomorphic sarcoma. It has better prognosis than other high grade sarcoma with recurrence rate of 40-75% & mortality rate of 28%.[3]

Based on the degree of differentiation, it is now a days classified into high & low grade. High grade de-differentiated liposarcoma is a cellular tumor with severe pleomorphism seen in non lipogenic component. Mitotic rate of > 5/10 hpf is considered significant.[4] High grade non-lipogenic component often resembles undifferentiated pleomorphic sarcoma whereas low grade non-lipogenic component in low grade de-differentiated liposarcoma resembles fibromatosis or well differentiated liposarcoma. De-differentiated liposarcoma are more sensitive for MDM2 & CDK4 immunostains compared to well differentiated liposarcoma.[5]

In addition, it is also positive for p16, Vimentin, p53, Rb and PPAR gamma.[6] Different case reports have presented varied clinical presentation and laboratory findings, but the diagnosis was confirmed using IHC with MDM2 and CDK4 positivity in majority of the cases (Table-1) [7-10].

It is essential to properly do sub-typing of sarcoma in order to decide the definitive line of treatment. In this case, based on histology, diagnosis of GIST and leiomyosarcoma were considered initially, but IHC confirmed the diagnosis and changed the line of treatment. So, it's crucial to do IHC to diagnose and plan treatment of sarcoma correctly.

Generally it can be treated using both chemotherapy & surgery if the mass is resectable with negative margins. [7-10] Chemotherapy was the treatment of choice in our case as surgery was not an option due to its unresectability.

Table 1 Showing comparison between different cases reports about their presentation and management

	Site	Clinical features	Laboratory findings	Immunohistochemistry	Treatment
Park et al[7]	Mesentry of ascending colon	Palpable abdominal mass	Within normal limits	MDM2, CDK4, Ckit positive	Surgery followed by chemotherapy
Cha et al[8]	Mesentry of small bowel	Palpable abdominal mass & urine frequency	Within normal limits including CA125, CEA, CA19.9	S100 positive	Surgery followed by chemotherapy
Dhakal et al[9]	Mesentry of small bowel	Pain & palpable abdominal mass	Within normal limits including CEA	S100, MDM2, CDK4 positive	Surgery followed by chemotherapy
Sawayam et al[10]	Ascending colon	Palpable mass	Within normal limits	MDM2,CDK, p16 positive	Only surgery
Our case	Recto-sigmoid colon serosa	Constipation, abdominal distension	CA125 raised significantly	MDM2,CDK, S100 positive	Only chemotherapy

Conclusion

De-differentiated liposarcoma of peritoneum is a rare malignancy with poor prognosis. Any tumor showing high grade or low grade non-lipogenic sarcoma component, one must keep this entity in the differential diagnosis. Immunohistochemistry must be used to do correct sub-typing of sarcoma so as to decide proper management which will be beneficial to the patient.

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