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Case Report

A rare case of retroperitoneal leiomyosarcoma

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ABSTRACT

Leiomyosarcoma, a rare malignancy of smooth muscle may arise from the retroperitoneum. They often reach a large size before diagnosis is made. Patient presents with vague symptoms, as such retroperitoneal malignancies are related to displacement of organs and obstructive phenomenon. The present paper is one of the very few case reports of retroperitoneal leiomyosarcoma which illustrates the presenting symptoms, gross and microscopic findings, treatment modalities and prognostic indicators of a 70 years old male seen at Government medical college, New Civil Hospital, Surat.

Keywords: Retroperitoneum, Leiomyosarcoma, Soft tissue sarcoma

INTRODUCTION

Soft tissue sarcomas constitute a heterogenous group of neoplasms of various histologies and comprise <1% of all adult malignancies. They occur at all anatomic body sites and about 10-20% of soft tissue sarcomas arise from the retroperitoneum.[1] Liposarcomas, pleomorphic sarcomas NOS and leiomyosarcomas are most common histologic types of retroperitoneal sarcomas. Leiomyosarcomas of retroperitoneum and abdominal cavity are one of the most common subgroup and are associated with an aggressive clinical course.[2] Leiomyosarcomas of somatic soft tissue are second but less common subgroup associated with better prognosis. Cutaneous leiomyosarcomas comprise a third subgroup which has a good prognosis. The last subgroup comprises leiomyosarcoma of vascular origin. Leiomyosarcomas may occur in an unusual soft tissue site such as head and neck and paratesticular region but these are uncommon.[2] The retroperitoneum provides an expansible anatomic location for tumor arising there, and these tumors often become very large before symptoms manifest.[1] Complete tumor resection is the treatment of choice but is rarely feasible. Although adjuvant chemo-radio therapy is associated with reduced risk of local recurrence, there has generally been no impact on survival. Another key factor related to prognosis is the grade of the tumor.

CASE REPORT

A 70 years old muslim male was admitted to our surgical department with a 3 months history of increasing abdominal girth, intermittent abdominal discomfort, a palpable abdominal mass and 8kg weight loss. Physical examination revealed a 25cm palpable mass over the left middle portion of the abdomen. Abdominal X-ray revealed a displaced transverse colon and a soft tissue density in the left middle portion of abdomen. Computed Tomography (CT) scan of abdomen also revealed an approximately 25x10x6 cm^3 sized, heterogenous retroperitoneal mass with contrast enhancement which could not be differentiated from the left kidney. Whole body CT scan/ USG did not reveal any metastatic lesion at any other site of the body. He underwent exploratory laparotomy with resection of 500gm retroperitoneal mass along with left kidney. In our department of pathology, we received a soft tissue mass in 2 parts, one measuring 10x6x6 cm^3 in size and another one measuring 11x10x5 cm^3 in size with attached kidney measuring 7.5x2.5x2 cm^3 in size. Mass was well circumscribed, however at

places, nodularity was seen. The mass appeared to be obliterating the left kidney. On cut section, mass was whitish in colour with greyish areas. On cut section of kidney, it showed normal appearing cortex; renal pelvis and medulla was obliterated by mass. Grossly, ureter was not identified.

Microscopic examination revealed high grade leiomyosarcoma with high cellularity, pleomorphic spindle shaped stromal cells arranged in interlacing fascicles and focally showing nuclear pallisading, about 4 mitoses/10hpf, foci of hyaline degeneration and coagulative necrosis (<50%) along with interspersed mononuclear infiltrate. Tumor cells exhibited positive cytoplasmic staining for Smooth Muscle Actin, Desmin and Vimentin and were immuno negative for S-100, EMA, CK, CD 117 (C-Kit) and Alk-1. The patient did not receive any chemo/ radio therapy and he is at present alive.

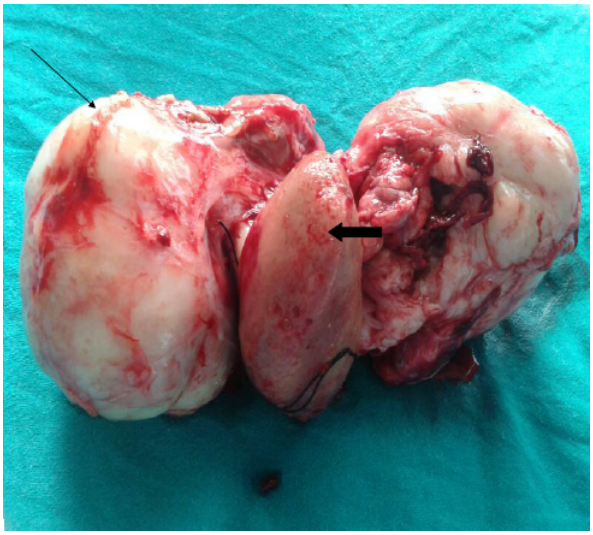

Figure 1: retroperitoneal mass (⟶) along with left kidney (➡).

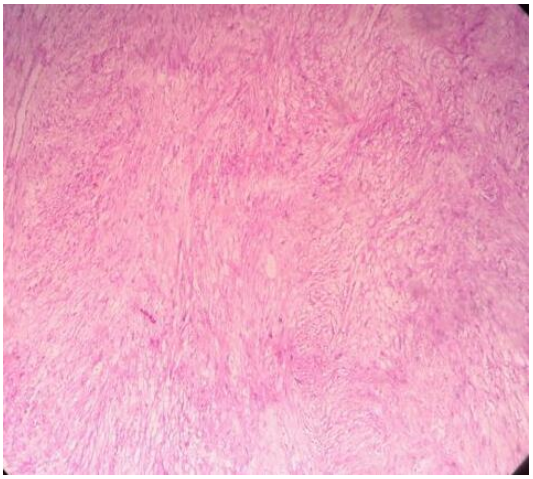

Figure 2: Smooth muscle cells arranged in interlacing fascicles (H & E stain; 10x view).

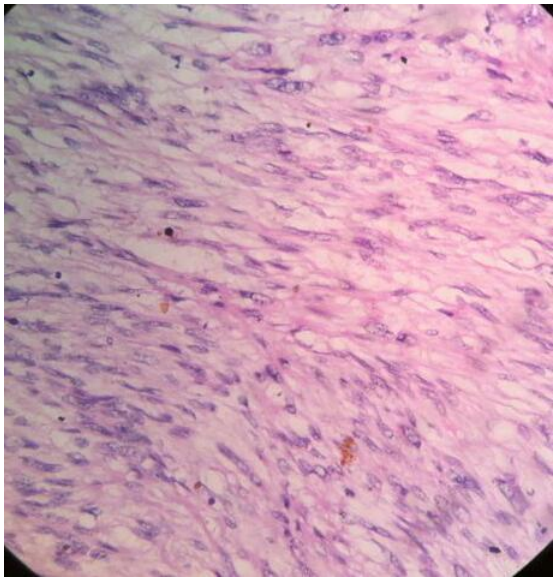

Figure 3: Pleomorphic spindle shaped cells with interspersed mononuclear infiltrate (H & E stain; 40 x view).

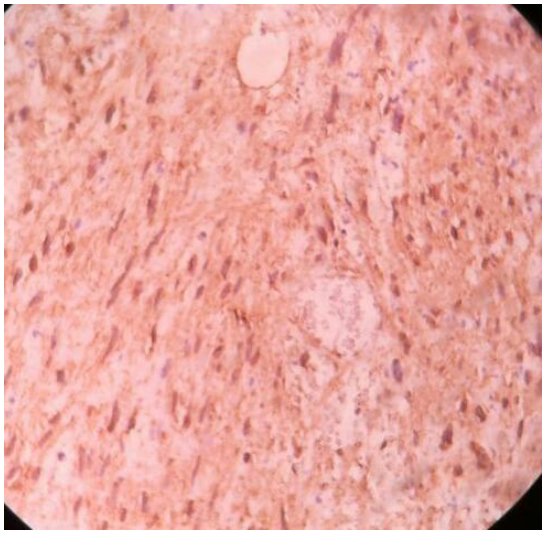

Figure 4: cells showing immunoreactivity for SMA (IHC; 40x view).

DISCUSSION

The microscopic pattern in our case raised a suspicion of other such tumors. Thus, the differential diagnoses put forth by us were benign counterpart of Leiomyosarcoma i.e. Leiomyoma, Extraintestinal GIST, Inflammatory Myofibroblastic Tumor and Malignant Peripheral Nerve Sheath Tumor.

The differential diagnosis between leiomyoma and leiomyosarcoma of soft tissues depends upon a combination of gross and microscopic features.[3] Size, cellularity, atypia, necrosis and mitoses per high power field are characteristics that differentiate between leiomyoma and leiomyosarcoma. Of these points,

mitoses/hpf is considered most reliable.[4] High mitotic activity (>5/50hpf), as in our case, is virtually diagnostic of malignancy. A diagnosis of leiomyosarcoma was also strongly suspected as the tumor was overly large (>10cm), necrotic and hemorrhagic.[3]

Extraintestinal GIST with a spindled pattern more closely resembles leiomyosarcoma but this case was distinguished from it by the fact that the cells in it usually have a short fusiform shape in contrast to elongated cells of leiomyosarcoma. They have an oval, centrally placed nucleus and lightly staining cytoplasm[2] and presence of cytoplasmic vacuoles at both the ends of nucleus.[3] The neoplastic cells are not arranged in long well-oriented fascicles like leiomyosarcoma but consist of short ill-defined fascicles, sometimes in a storiform pattern.[2] On IHC, nearly all GISTs are CD117 positive, making this antigen the most sensitive and specific means of confirming the diagnosis. One-half to two-thirds of GISTs are CD34 positive and one quarter, SMA positive. Desmin and keratin are rarely present (<5%).[2] h-caldesmon is frequently present and myosin is erratically present.[3]

Malignant Peripheral Nerve Sheath Tumor (MPNST) shows presence of serpentine shape of tumor cells, arrangement in palisades or whorls, marked contrast between the deeply hyperchromatic nuclei and pale cytoplasm (punched out nuclei), in contrast to elongated blunt ended nuclei and acidophilic fibrillary cytoplasm of cells in leiomyosarcoma. They have perivascular concentration of tumor cells with plumper shape and geographic areas of necrosis with tumor pallisading at edges. Mitoses are usually abundant.[3] On IHC, tumor cells show reactivity for Schwann cell markers such as S-100 and Leu7 in approximately half of the cases.[3]

One of the pattern of Inflammatory Myofibroblastic Tumor (IMT) which is characterized by a compact fascicular spindle cell proliferation with variable myxoid and collagenized regions and distinctive inflammatory infiltrate, small aggregates of plasma cells or lymphoid nodules also resembles a leiomyosarcoma.[5] However, on IHC, it shows Cytoplasmic positivity for Alk-1 in approximately 50% of cases, strong and diffuse cytoplasmic reactivity for vimentin (in virtually all IMT), reactivity for SMA and muscle specific actin varying from focal to diffuse pattern in spindle cell cytoplasm. Desmin is identified in many cases. Focal CK is seen in one third of cases. Myogenin, myoglobin, CD 117 and S-100 are negative.[5]

Leiomyosarcoma on the other hand shows immunoreactivity for Vimentin, Smooth Muscle Actin, Desmin, h-caldesmon and basal lamina components.[3] Positivity for atleast 2 of these markers is more supportive of leiomyosarcoma.[5] S-100, EMA,CK, C-Kit (CD 117) and Alk-1 are usually negative.[5] Thus, the Immunohistochemistry features also in our case favour the diagnosis of leiomyosarcoma and rule out the other differentials.

CONCLUSION

Retroperitoneal Leiomyosarcoma is a rare malignancy. It may present in fifth to seventh decade with vague symptoms. Thus any tumor with such presentation having microscopic finding of cells with elongated blunt ended nuclei arranged in bundles should raise a suspicion of retroperitoneal leiomyosarcoma. Early recognition and aggressive surgery are keys to long term survival but the mainstay of treating advanced disease is yet to be determined and needs further research as the overall prognosis of advanced disease is relatively poor.

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