Primary retroperitoneal inflammatory leiomyosarcoma with metaplastic bone formation: Report of a rare variant with a brief review of literature

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Abstract:
Retroperitoneal sarcomas constitute a heterogenous group of neoplasms and often pose a diagnostic challenge. Leiomyosarcomas are a common subgroup, however improved diagnostic techniques, in particular immunohistochemistry, electron microscopy and cytogenetics, have provided a new insight into tumours in which the lines of histogenesis or differentiation were previously unknown or only speculative. We present a case of a retroperitoneal sarcoma in a 57 year old male patient, which after complete work up was diagnosed as inflammatory leiomyosarcoma. There was a focus of heterologous bone formation also in our case, which has not been reported till date. The documentation of this entity is not only very limited in literature, but is also important as the prognosis of the completely resected tumour is better than other variants in this group.

Key words: Immunohistochemistry; Inflammatory leiomyosarcoma; Retroperitoneal; Metaplastic bone; Sarcoma

Introduction
Sarcomas are rare malignant tumours that arise from mesenchymal tissue at any site of the body and constitute 0.7% of adult malignancies. Ten to twenty percent of soft tissue tumours occur in the retroperitoneum as it provides a widely expansile anatomic location for tumours arising there and which often attain a large size before symptoms manifest [1]. Although leiomyosarcomas are the second most common category of sarcomas in this region, the inflammatory variant is a very rare entity, which was classified originally within the malignant
fibrous histiocytoma group with which it shared many similar histological features.

**Case Presentation**

A 57 year old poorly built man, presented to the surgical outpatient department with complaints of generalized weakness and abdominal pain since a month. Physical Examination revealed a 14 cms palpable mass, in the mid-abdominal region. Abdominal computed tomography showed a heterogeneous mass in the retro peritoneum, involving a portion of the pancreas. A CT Guided FNAC was done which showed features of a soft tissue sarcoma of intermediate grade. Hemoglobin was 10 gm%, all other biochemical parameters were within normal limits. The patient was taken up for exploratory laparotomy, and a multilobulated grey white mass was debulked in two pieces, with a 2.5 cms wide clearance margin, with a portion of the involved pancreas being removed, altogether weighing 435 gms. Gross examination of the specimen showed two grey white irregular masses, larger being 10 x 8 x 3 cms. Cut section of both the masses showed homogeneous white areas, at places appearing whorled, with areas of softening.

Microscopic examination showed a highly cellular malignant spindle cell neoplasm with cells with cigar shaped nuclei arranged in fascicles and whorls, with few foci showing a storiform pattern. Also noted were areas of xanthomatous cells, tumour giant cells, tumour necrosis. Two additional features were prominent areas of inflammatory cells, mainly lymphocytes and a focus of well formed mature bone.

Figure: 2a 2b, 2c and 2d :
Immunohistochemistry revealed diffuse positivity for desmin, smooth muscle actin, HHF-35 and calponin. S-100, MDM-2, caldesmon were negative. CD68 was positive in the foamy cells, with spindle cells being invariably negative for this marker.

After literature search, a final diagnosis of primary high grade inflammatory leiomyosarcoma of the retroperitoneum was made. The patient received chemotherapy and is without recurrences after a follow up of till date.

**Discussion**

The histogenesis of retroperitoneal tumours and its treatment underwent evolution since they were described in 1829 by Lobstein. Liposarcomas are the most common retroperitoneal sarcomas, leiomyosarcomas being the next common type. They most commonly occur in the 5th to 7th decade, with no predilection for gender [2].

These tumours have vague presenting symptoms, with the most common complaint being back pain, weight loss or increasing abdominal girth. Many a times masses are discovered incidentally, during routine examination or abdominal surgery for other reasons [2,3].

As the retroperitoneal area is not easily accessible to palpation, the diagnosis is often delayed because of its asymptomatic nature in an early stage. Imaging studies contribute greatly to delineate the size, location and characterization of the mass, with abdominal CT being the preferred imaging tool. A leiomyosarcoma on CT and MRI is typically a non-fatty, irregularly margined, heterogeneous mass without calcification. Ultrasound or angiography may be useful in delineating tumour vessels or vascular infiltration if these are suspected [1].

Typical Lieomyosarcomas on microscopy show characteristic features like fascicles and bundles of spindle cells, with cigar shaped nuclei,
plenty of mitotic figures, tumour giant cells and areas of necrosis. However, in 1995 a variant called inflammatory leiomyosarcoma was first described, with a morphology similar to that of conventional leiomyosarcoma but with additional features of xanthoma cells and prominent inflammatory infiltrate, mainly lymphocytes [4,5]. Few well recognized tumours have significant inflammatory component, the sarcoma type being inflammatory variant of well differentiated liposarcoma, which was ruled out in our case as lipoblasts were not seen. The implications of this previously uncharacterized variant of leiomyosarcoma lie mainly with the tumour type, inflammatory Malignant Fibrous Histiocytoma [MFH], with which it shares many morphological features [6,7]. Finding of mitotic activity within the xanthomatous cells, more of a storiform pattern of spindle cells and a prominent admixture of most often acute inflammatory cells favor MFH [7].

While these tumours express desmin to a certain degree, they lack or only focally express other markers including muscle specific actin, smooth muscle actin [SMA] and Caldesmon, leading to a recent suggestion that they may not be true smooth muscle tumours. The xanthomatous cells are known to express CD 68, as in our case and can be a finding in a variety of mesenchymal tumours including smooth muscle neoplasms [4].

In the literature search of this entity nowhere did we come across bone formation in the tumour, which was seen in our case. Although foci of high pleomorphism were found in our case, areas histologically typical of leiomyosarcoma and positivity of smooth muscle markers even in these areas aided the diagnosis [8]. The bony foci did not stain for the smooth muscle markers, suggesting that it is a heterologous entity. The bone formation was benign, of metaplastic nature, characterized by bony trabeculae rimmed by benign looking osteoblasts. No lipoblasts were identified in the sections. Also, MDM-2 and S-100 were done in our case, both of which were negative to rule out dedifferentiated liposarcoma and MFH, which may show foci of metaplastic bone formation [9-11].

Cytogenetically also hyperhaploidy is a distinguishing characteristic of inflammatory leiomyosarcomas along with disomy for some chromosomes, notably 5 and 20. This feature is unlike other sarcomas, which otherwise have a low chromosome number [12].

Complete surgical resection with at least 3 cms of tumour free margins is the treatment of choice, but is rarely feasible due to invasion of adjacent structures by the tumour. Although initially associated with very good prognosis, recent cases have been reported with recurrences and metastasis, the cause being inadequate surgical removal [13]. Chemotherapy is not effective and radiation is limited by toxicity to the adjacent structures [14]. Factors which influence the outcome include size, grade, and weather extension to bone and nerve is present. Nevertheless, prognosis of inflammatory leiomyosarcoma seems to be favourable when treated early with wide excision, unlike other retroperitoneal sarcomas like typical or dedifferentiated leiomyosarcomas, liposarcomas and MFH’S which exhibit worst clinical behavior [14,15].

**Conclusion**

We have reported a rare case of retroperitoneal inflammatory leiomyosarcoma. Not only the documentation of this is very limited but also our case had a focus of heterologous mature bone formation, which has not been reported till date. The clinicians must be aware of this entity as it has a relatively better prognosis than other sarcoma variants in the retroperitoneal region if treated with wide excision.

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