

Primary leiomyosarcoma of peritoneal cavity

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Abstract

Leiomyosarcomas of soft tissue are the rare tumors and the retroperitoneum is the most common site involved. We report a case of primary leiomyosarcoma of the peritoneal cavity which clinically presented with suprapubic, freely mobile, nontender mass which measured 10×10 cm in size. Contrast enhanced computed tomography revealed well defined heterogeneous hypodense solid cystic mass. The mass was surgically excised out in its entirety. The histopathological examination revealed spindle cells arranged in alternating fascicles having pleomorphic nuclei, indistinct margin and eosinophilic cytoplasm with foci of haemorrhage, necrosis and 5-6 mitosis/HPF. The spindle cells were immunoreactive for smooth muscle actin, desmin and negative for S-100, CD-34 and c-kit. Histopathology and immunohistochemistry were helpful in making the final confirmatory diagnosis. Leiomyosarcomas are aggressive tumors, with poor prognosis and often difficult to treat. The survival rates are lowest among all soft tissue sarcomas.

Introduction

Leiomyosarcomas of soft tissue are rare tumors that account for only 10% of soft tissue sarcomas and the most common site for leiomyosarcoma of soft tissue is the retroperitoneum, accounting 50% of all the cases.¹ The precise diagnosis based on imaging alone is difficult and the final diagnosis is obtained after histopathological examination. Leiomyosarcoma presentation as diffuse peritoneal or omental mass, however, is very rare.

We report a case of primary leiomyosarcoma of peritoneal cavity which was treated with surgical excision.

Case Report

A 65-year-old female presented with lower abdominal lump and pain for 3 months. On local examination the mass was suprapubic, freely mobile, nontender and measured 10×10

cm in size. contrast enhanced computed tomography revealed well defined heterogeneous hypo dense solid cystic mass approx. 12×10×8 cm in peritoneum with area of calcification, necrosis and multiple septation (Figure 1). Tumor marker (CEA: 1.9 ng/mL, CA125: 6.064 U/mL) were within normal limits.

The mass was surgically excised out in its entirety and peroperative finding revealed unremarkable adnexa and soft tissue mass arising from the mesentery. Grossly globular grey white soft tissue received which measured 15×10×7 in size. The cut section was solid, cystic with large area of haemorrhage and necrosis. Multiple histopathological sections examined revealed spindle cells arranged in alternating fascicles having pleomorphic nuclei, indistinct margin, eosinophilic cytoplasm with foci of haemorrhage and necrosis. Mitotic figures were 5-6/HPF (Figure 2A-C). The tumor cells were immunoreactive with smooth muscle actin (Figure 2D), desmin and negative for S-100, CD-34 and c-kit. There was no evidence of lung, liver or nodal metastases and peritoneal seeding. Final diagnosis of primary leiomyosarcoma of peritoneal cavity was made. The postoperative course was uneventful; however the patient expired after six month of surgery.

Discussion

The primary peritoneal tumors arise in the peritoneum in absence of a visceral site involvement and they mainly include mesothelioma, serous carcinoma, leiomyomatosis peritonealis disseminata, desmoplastic small round cell tumor and solitary fibrous tumor. The metastatic neoplasms in peritoneum are lymphomatosis, pseudomyxoma peritonei and carcinomatosis secondary to ovarian and gastrointestinal neoplasm. The infection which form the peritoneal and omental mass are disseminated histoplasmosis and tuberculosis.² Soft tissue leiomyosarcoma has classically been subdivided into three groups for prognostic and treatment purposes which includes leiomyosarcoma of somatic soft tissue, cutaneous leiomyosarcoma and leiomyosarcoma of vascular origin. Leiomyosarcoma are more common in female than males with poor prognosis and aggressive clinical course as seen in our case.¹ Leiomyosarcoma spread into peritoneal cavity by embolic metastases or when tumor grows toward the subserosa from its usual site of origin in the muscle layer or it may undergo central excavation and perforate into the peritoneal cavity.^{3,4} The CT findings of peritoneal leiomyosarcoma closely resemble rare, benign condition of leiomyomatosis peritonealis disseminata which is commonly associated with uterine leiomyoma in the form of multiple small

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nodules of smooth muscle cells throughout the subperitoneal surfaces,⁵ or those of intraperitoneal drop metastases from hepatocellular carcinoma because of the similar way of intraperitoneal spread.⁶ The histological features under light microscopy are the most important factors in making the diagnosis of leiomyosarcoma, immunohistochemistry can be extremely useful when the morphologic findings alone are insufficient for a definitive diagnosis, vimentin will be consistently expressed by leiomyosarcomas and desrnin expression are focal or may be absent. Leiomyosarcomas show variable expression of smooth muscle actins.⁷ Tumors with 1-4 mitoses per 10 HPF should be regarded as potentially malignant, and those with 5 or more mitoses per 10 HPF as malignant.⁸ The present case showed mitotic activity of 5-6/ HPF. The staging of leiomyosarcoma is important in guiding treatment and providing prognostic information. The AJCC system classifies the tumor based upon histologic grade high or low, the tumor size more or less than 5 cm, location as superficial or depend the presence or absence of metastatic disease.⁹ Overall reported survival for patients diagnosed with soft tissue leiomyosarcoma range from 50% of 3-year survival to 64% of 5-year survival, making this tumor one of the more aggressive soft tissue sarcomas. The prognosis is poor, with survival rates among the lowest of all soft tissue sarcoma.¹⁰ The local control is obtained with wide surgical excision and neoadjuvant or adjuvant radiation therapy whenever it is appropriate where local control is an issue and chemotherapy is employed for the treatment of systemic disease.¹⁰

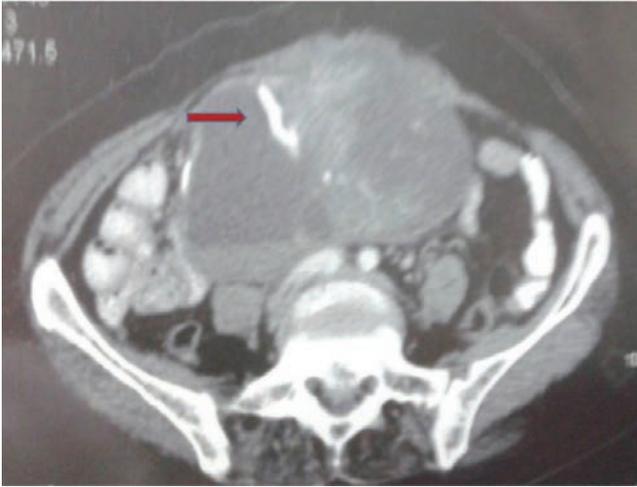


Figure 1. Computed tomography abdomen scan showed well defined approx 12×8×10 cm heterogeneously hypodense mixed solid and cystic mass (arrow) in the abdominal cavity.

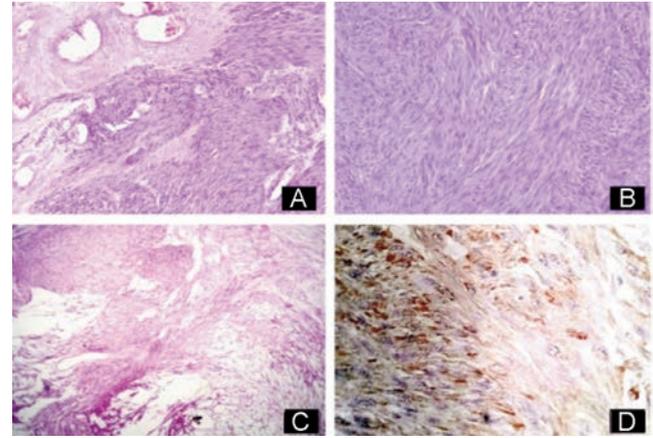


Figure 2. A) Tumor cells admixed with foci of haemorrhage and necrosis (×100). B) Section shows spindle cells arranged in alternating fascicles with pleomorphic nuclei and indistinct margin (×400). C) The tumor cells are entrapping the adipose tissue (×100). D) The tumor cells were immunoreactive with smooth muscle actin (×600).

Conclusions

Leiomyosarcomas is rare soft tissue sarcomas and this case also adds to the existing small list of similar uncommon tumor of this type. The histopathological and immunohistochemical examination are helpful in making the final confirmatory diagnosis.

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