International Journal of Scientific Reports
Kumar V et al. Int J Sci Rep. 2024 Dec;10(12):447-450
http://www.sci-rep.com
pISSN 2454-2156 | eISSN 2454-2164
DOI: https://dx.doi.org/10.18203/issn.2454-2156.IntJSciRep20243495

Case Report

Massive retroperitoneal leiomyosarcoma: a case report

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Received: 07 August 2024
Revised: 26 October 2024
Accepted: 16 November 2024

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ABSTRACT

Soft tissue sarcoma is mesenchymal tumor of malignant in nature seen in body. Liposarcoma followed by leiomyosarcoma are most common type. Leiomyosarcoma (LMS) in retroperitoneum region develop from smooth muscle cell or from wall of large vein and detect clinically when attain huge size, causing symptom due to compression or displacement to surrounding structure. We are presenting case report of a 65 years old lady presented with progressive pain abdomen, distension, loss of weight and appetite for 1 year. Radiological investigation reveals a large bilobed intra-abdominal mass with no major vascular involvement. Patient underwent complete surgical excision with resection of surrounding structure and post operative period was uneventful. HPE and IHC of resected tumor were suggestive of Leiomyosarcoma. Leiomyosarcoma usually has slow and silent progression hence present clinically when compression or displacement of adjacent structure arises. It has malignant potential. Radiological investigation helps in deciding resection. R0 resection carries very good prognosis since chemo and radiotherapy has limited role in complete cure.

Keyword: Retroperitoneal leiomyosarcoma, Soft tissue sarcoma, Liposarcoma

INTRODUCTION

Soft tissue sarcomas are malignant mesenchymal neoplasms representing less than 1% of cancers in adults and it affect peritoneum in 12 to 69% of the cases.[1] Liposarcoma is the most common type while leiomyosarcoma is the second most common variant.

Leiomyosarcoma is seen between 54 to 65 years mainly affect female more than male.[2] Leiomyosarcoma (LMS) develops from retroperitoneal smooth muscle cells or from cells in the walls of large peritoneal veins, presenting macroscopic appearance with gray, white, or brownish coloring, and may also cause hemorrhage, necrosis, and cystic changes when they reach large sizes. Generally, the mass has well circumscribed margins, with regions of infiltration into adjacent tissues.[2,3]

CASE REPORT

A 65 years old lady with no known co-morbidity present to our tertiary health care Lady Hardinge Medical College and associated Shrimati Sucheta Kriplani hospital with chief complains of gradually progressive abdomen distension, pain in abdomen, loss of appetite and loss of weight for 1 year. On examination a large bilobed, bosselated, intraabdominal mass of size about 30×20 cm was present occupying almost whole of abdomen and extending into the pelvis with an irregular surface, not moving with respiration, restricted mobility, firm to hard in consistency, dull on percussion, no evidence of free fluid and no other lump was present. (Figure 1) CECT shows a large right sided heterogeneously enhancing mass lesion approximately 27.5×13.7×16.8 cm (CC X TR X AP) in retroperitoneal

region displacing bowel loops and aorta to left. Right kidney showing hydronephrosis and proximal hydroureter. Left kidney and ureter was unremarkable. Coronal section image showing the extent of the retroperitoneal mass extending from subhepatic region to deep into the pelvis. Mass was not involving major vasculature (Figure 2).

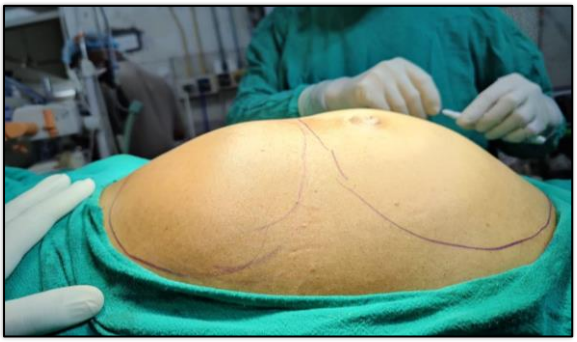

Figure 1: Clinical presentation of intra-abdominal mass surface marking.

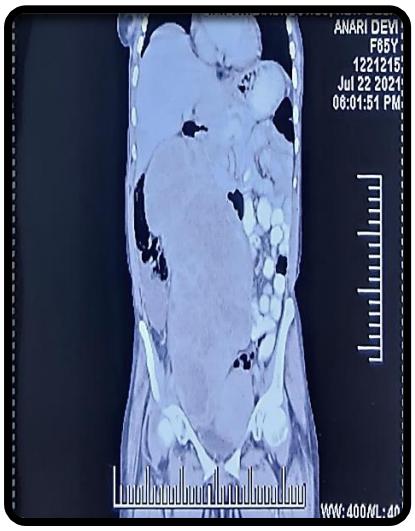

Figure 2: Extent of mass (coronal section) on CECT.

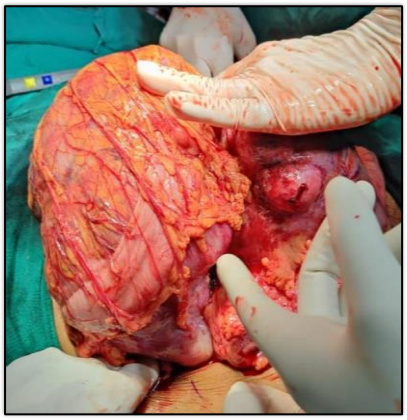

Figure 3: Intra-abdominal mass adhered to small and large intestine.

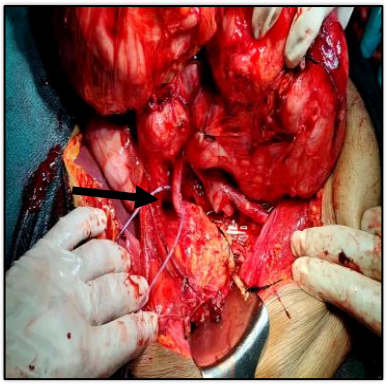

Figure 4: Right mid ureter involvement (arrow).

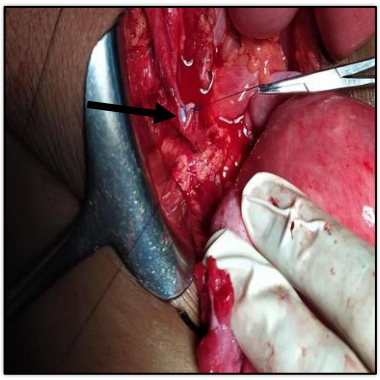

Figure 5: Uretero-ureteric anastomosis over DJ (arrow).

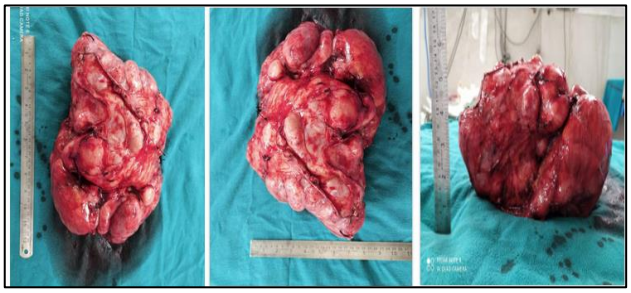

Figure 6: Dimensions of resected specimen (30×22×14 cm).

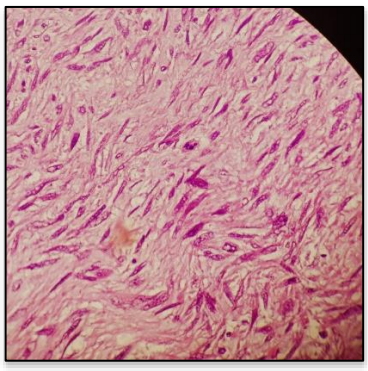

Figure 7: H & E staining with atypical mitoses.

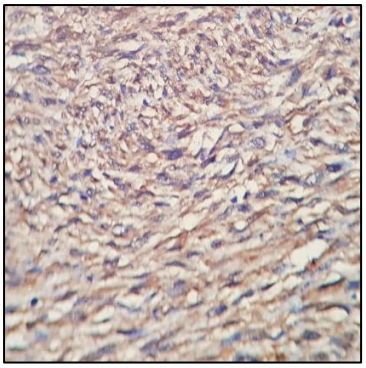

Figure 8: Smooth muscle actin (positive).

Operative procedure

Intraoperatively there was large bosselated retroperitoneal mass arising from right perinephric region and adhere with mesentery of terminal ileum, caecum, ascending colon, proximal 1/3rd of transverse colon and rest of bowel was pushed toward left of peritoneal cavity (Figure 3). Mild amount of hemorrhagic ascitic fluid seen. The mass was encasing the midpart of right ureter causing proximal right hydro-uretero-nephrosis (Figure 4). And no major vascular involvement was seen.

Subsequently patient underwent complete excision of retroperitoneal mass with right extended hemicolectomy with resection of involved right ureter. Gastrointestinal continuity was done by side-to-side ileo-transverse anastomosis and right urinary continuity by end-to-end uretero-ureteric anastomosis over double-J stent (Figure 5) with subhepatic and pelvic drain placement under general anesthesia. Resected gross specimen was bilobed in shape and weighing approximately 12 kg and was measuring 30×22×14 cm in dimensions (Figure 6). Postoperative course was uneventful and the patient was discharged on the 10th postoperative day. Histopathology examinations showed atypical mitosis on H&E staining (Figure 7), SMA positive (Figure 8), Desmin positive and vimentin positive, all feature consistent with leiomyosarcoma. Last follow up, 3 months after surgical resection, the patient had no complaints and there was no recurrence of retroperitoneal leiomyosarcoma on radiological evaluation.

DISCUSSION

Leiomyosarcoma (LMS) are stromal tumor with smooth muscle differentiation and malignant in nature. The incidence of soft tissue LMS increases with age, reaching its peak in the 6th to 7th decade of life and affecting women more often. In histology LMS characterize by spindle cells forming fascicles with well-defined margins, and the presence of at least one of the following criteria for diagnosis: cellular pleomorphism or atypia, coagulative tumor necrosis, or over 10 mitotic figures per 50 high-power fields in women, and over 1 mitotic figure per 50 high-power fields in men.[2] LMS mainly arise from retroperitoneum region, lower limbs and solid organs.[4] LMS present with symptoms due to mass compressing or displacing adjacent structure. Compression on inferior vena cava at lower portion can lead to lower limb edema, compression or obstruction of the hepatic veins in the upper portion can result in Budd-Chiari syndrome, characterized by hepatomegaly, jaundice, and ascites. Bowel displacement leads to chronic constipation or recurrent attacks of bowel obstruction. Radiological studies i.e., MRI and contrast-enhanced CT lack specificity but help in delineating the tumor's relationship with adjacent structures and vessel involvement and resectability.[5]

Predisposing factors contributing to its development includes Li-Fraumeni syndrome, hereditary retinoblastoma, and exposure to radiation.[6] On IHC, at least one myogenic immunomarker such as smooth muscle actin, desmin, or h-caldesmon shows positivity in 100% of the cases, with over 70% demonstrating positivity for multiple markers. Since none of these markers are entirely specific for smooth muscle differentiation, positivity for two markers proves more conducive for an accurate diagnosis.[7] Surgical resection with tumor-free margins is considered the primary treatment for retroperitoneal sarcomas but due to late presentation and diagnosis tumor invade adjacent structure and R0 resection become very challenging which leads to recurrences and mortality. Thus, en bloc resection of adjacent organs is the recommended therapy according to the guidelines of the European Society for Medical Oncology for good prognosis.[8] The combination of gemcitabine and dacarbazine is one of the chemotherapy options that has shown positive results in LMS. The main limitation of the study is, single case report, which limits the wider application of this study. Studies on larger patient groups are required to validate the results.

CONCLUSION

Soft tissue retroperitoneal leiomyosarcoma is 2nd most common type of retroperitoneal sarcoma. It usually has slow and silent progression hence present clinically when compression or displacement of adjacent structure arises. It has malignant potential. Radiological investigation helps in deciding resection. R0 resection carries very good prognosis since chemo and radiotherapy has limited role in complete cure.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Kumar V, Pathania P, Siddiqui AA, Kumar D, Jain R. Massive retroperitoneal leiomyosarcoma: a case report. Int J Sci Rep 2024;10(12):447-50.