Content available at: https://www.ipinnovative.com/open-access-journals

IP Indian Journal of Anatomy and Surgery of Head, Neck and Brain

Journal homepage: https://www.ijashnb.org/

Case Report

Myxofibrosarcoma (A rare retroperitoneal tumour)-A case report

Supriya[1], Vijay Verma[2,*], Anupam Nanda[1]

[1]*Dept. of General Surgery, Dr. Rajendra Prasad Government Medical College, Tanda, Kangra, Himachal Pradesh, India*
[2]*Dept. of General Surgery, Indira Gandhi Medical College & Hospital, Shimla, Himachal Pradesh, India*

ARTICLE INFO

Article history:
Received 20-03-2021
Accepted 20-03-2021
Available online 21-04-2021

Keywords:
Soft tissue sarcoma
Retroperitoneal tumour
Myxofibrosarcoma.

ABSTRACT

This is a case report that discusses about a rare retroperitoneal tumour (incidence- 0.3-3%). This case was admitted at Dr. RPGMC Tanda with the complaints of lump abdomen and was further diagnosed as myxofibrosarcoma.

1. Introduction

Soft tissue neoplasms are rare and unusual neoplasms, approximately 1% of all adult human cancers and 15% of all paediatric malignancies.[1] Their site of occurrence is in the following order: extremities (45%), viscera (20%), retroperitoneum (15%), thoracic (10%) and others (10%).[2]

Histological subtypes are classified as limited, intermediate and substantial or high metastatic potential. Limited metastatic potential subtypes include lipomatous tumour, dermatofibrosarcoma, hemangiopericytoma and desmoid tumour. Subtypes with intermediate metastatic potential include myxoid liposarcoma, myxoid MFH (malignant fibrous histiocytoma) and extraskeletal chondrosarcoma. Substantial/high metastatic potential subtypes include angiosarcoma, leiomyosarcoma, rhabdomyosarcoma, clear cell sarcoma, synovial sarcoma, pleomorphic sarcoma and dedifferentiated sarcoma.[3]

Most of the soft tissue sarcomas are thought to be sporadic and their cause is unknown. In rare cases, genetic factors, environmental factors, prior radiation therapy, viral infections and immunodeficiency have been associated with the development of sarcomas. Sarcomas have also been reported to arise in scar tissue, fracture sites or anatomic regions associated with prior soft tissue trauma. Genetic syndromes like Neurofibromatosis, Li-fraumeni syndrome, FAP (familial adenomatous polyposis) have all been shown to be associated with the development of soft tissue sarcomas.[2] A soft tissue sarcoma (STS) is one of the most common types of radiation associated tumors in the general population.[4] It is generally accepted that sarcomas are induced in heavily radiated tissues of the patients who have received 50Gy or more in or close to the radiation fields. The median interval between radiation exposure and the development of sarcoma is 10 years and this varies by histological type, with the shortest latency observed in liposarcoma (median 4.3 years) and the longest in leiomyosarcoma (median 23.5 years).[5]

High level occupational exposure to phenoxyacetic herbicides, chlorophenols and dioxins have also shown an increased incidence of soft tissue sarcomas (STS). However there is no positive correlation between dioxin concentration and STS. In fact, sarcoma risk was highest among those having the lowest dioxin level.[6,7]

The genomic alterations in STS are limited to only the most recurrent alterations. Heritable retinoblastoma gene (RB1) mutations are associated with an increased risk of

* *Corresponding author.*
E-mail address: docvjverma@gmail.com (V. Verma).

https://doi.org/10.18231/j.ijashnb.2021.007

bone and soft tissue sarcoma.[8]

Retroperitoneal sarcomas usually present as painless lump in the abdomen with no functional impact, although pain is noted in 33% of patients. Majority of them are asymptomatic. Patients with intra-abdominal or retroperitoneal sarcomas often experience nonspecific abdominal discomfort and gastrointestinal symptoms before diagnosis. However they can also present with gastrointestinal bleeding, incomplete obstruction and neurological symptoms.[9]

Here, we report one such case of soft tissue sarcoma whose site of origin was retroperitoneum. It presented as abdominal lump involving whole of the left side of the abdomen and it was further diagnosed histologically as myxofibrosarcoma.

2. Case Report

A 45 year old man presented to us with a lump in the left side of the abdomen for 2 months and generalized weakness for 1 month. There was no history of vomiting, anorexia or jaundice. There was a history of significant weight loss . Bladder and bowel habits of the patient were normal. On per abdominal examination, a lump of size 15x12cm was found occupying almost whole of the left side of abdomen (LHC,LL,LIF). The lump was found to be crossing midline and causing abdominal distension. It was firm in consistency, having ill defined margins and had smooth surface. The lump was found to be non pulsatile and non tender. There was no visible peristalsis. The lump was not moving with respiration. It was non ballotable. There was no hepatomegaly. Spleen could not be palpated. Clinically pallor was present. Digital rectal examination was grossly normal. Chest x ray was normal. CEA and CA19-9 was within normal limits. Colonoscopy was found to be a normal study. Peripheral smear showed a microcytic hypochromic picture. Iron studies like ferritin was normal and TIBC was low. Serum Fe was low. USG abdomen showed a large heterogenous mass of 15x14 cm likely origin from suprarenal region left side likely malignant. Contrast enhanced computerized tomography of the abdomen (CECT) showed a large rounded heterogenous lesion of size 18x17x28cm arising retroperitoneally on left side of abdomen in left pararenal space, with enhancing solid and fatty component, superiorly extending upto splenic hilum, medially crossing midline, displacing gutloops without any loss of fat planes, displacing pancreas, inferiorly extending upto pelvis with displacement of left kidney. No invasion of aorta or its branches or ivc was seen? liposarcoma ?? leiomyosarcoma. CECT of thorax had no evidence of metastasis.

2.1. Intra operative findings

A large retroperitoneal mass of size 35X20cm was found encasing spleen and left kidney, abutting splenic vein. Superiorly mass was extending upto post wall of stomach. En bloc resection of the mass along with left kidney and spleen was done. No ascites or peritoneal metastasis was found. The weight of the mass was found to be 5.4kg.

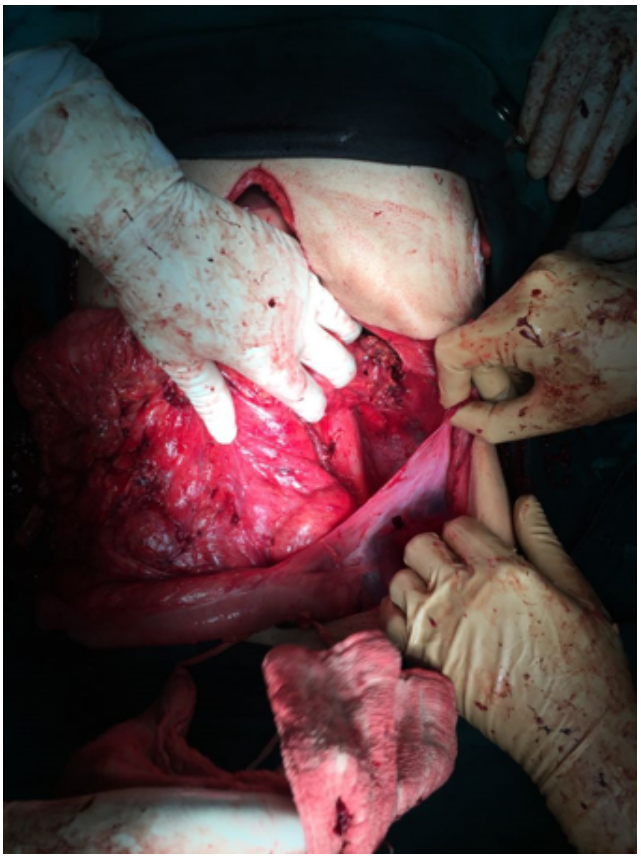

Fig. 1:

3. Discussion

Myxofibrosarcoma (MFS) is a variant of the group of malignant fibrous histiocytomas.[1] It is one of the most aggressive types of soft tissue neoplasms. It occurs mainly in people between 50-70 years of age and is more common in men than women. It exhibits a high local recurrence rate and a significant metastatic rate.[3] 5yr survival rate is generally 60-70%.[2] The histopathologic patterns of myxofibrosarcoma are characterized by a myxoid component of extracellular matrix, pleomorphic spindle cells, and curvilinear blood vessels. There are no specific immunohistochemical markers or genetic profiles for MFS, but the techniques are useful in excluding similar but differential tumors. Superficial MFSs often consist of multiple palpable nodules, while the deep-seated lesions

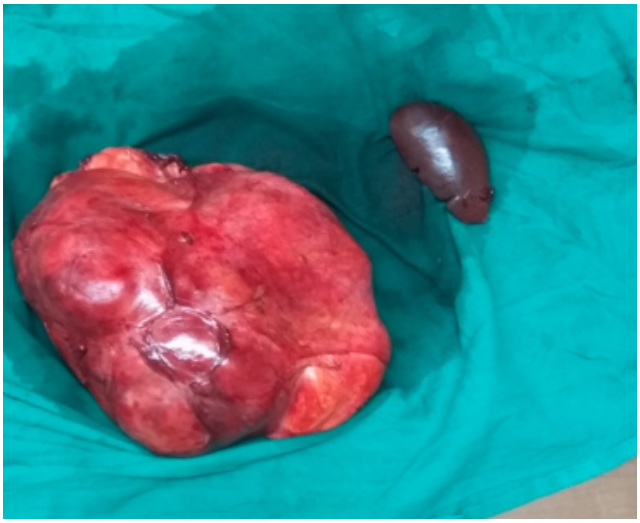

Fig. 2:

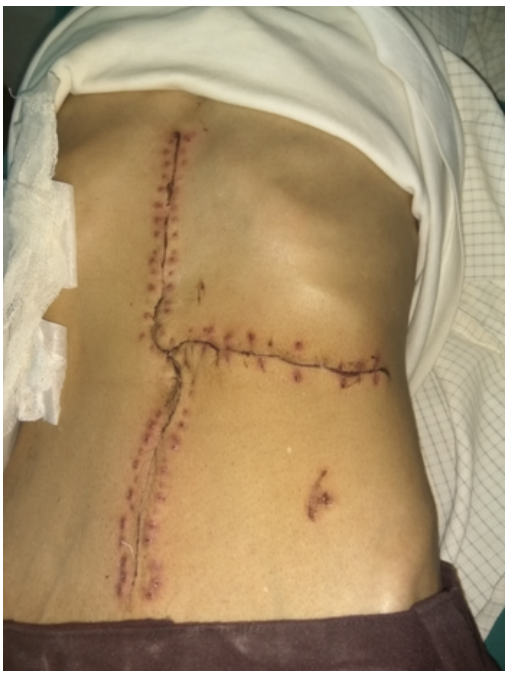

Fig. 3:

more often form a single mass. The tumors have a peripheral infiltrative growth pattern with extension along vascular and fascial planes extra or intramuscularly. [10]

Point of interest in this case was inspite of such a big mass enclosing both spleen and left kidney, there was no any evidence of metastasis, which is quite rare with such a large tumour.

Histopathology showed features of malignant mesenchymal tumour possibly myxofibrosarcoma. Sections from spleen and kidney was within normal histological limits.

On cross sectional examination of the specimen, there were multiple cystic and solid components with multiple dilated vessels within the mass.

4. Conclusion

The overall retroperitoneal tumour incidence is 0.3-3%.

The most common retroperitoneal tumour is liposarcoma (50%), followed by leiomyosarcoma (29%).

In children the most common retroperitoneal neoplasm is rhabdomyosarcoma.

Lymph node metastasis is rare (<5%). Sarcomas spreading to lymph nodes like rhabomyosarcoma, lymphangiosarcoma and epitheloid sarcoma, the local recurrence rate is 40-82%.

Retroperitoneal neoplasms are usually relatively chemoresistant tumours.

Typical chemotherapeutic regimes like AIM, MAID and AD are used in sarcomas. AIM consists of Adriamycin, ifosfamide and mesna. MAID regimen is a combination of mesna, doxorubicin, ifosfamide and dacarbazine. AD comprises of adriamycin and dacarbazine.

Radiation can be an effective treatment to decrease local recurrence of soft tissue sarcomas. The current National Comprehensive Cancer Network guidelines recommend radiotherapy for extremity sarcomas for high grade lesions, low grade lesions>5 cm or positive margins. [11]

These are also related to peripheral nervous system.

Our case finding was rare because inspite of having a large retroperitoneal tumor which is itself a less common neoplasm with a rare site of origin, there was no metastatic evidence.

5. Conflicts of Interest

All contributing authors declare no conflicts of interest.

6. Source of Funding

None.

References

1. Jemal A, Siegel R, Ward E, Hao Y, Xu J, Thun MJ, et al. Cancer Statistics, 2009. *CA: A Cancer J Clin*. 2009;59(4):225–9. doi:10.3322/caac.20006.
2. Malkin D, Li F, Strong L, Fraumeni J, Nelson C, Kim D, et al. Germ line p53 mutations in a familial syndrome of breast cancer, sarcomas, and other neoplasms. *Sci*. 1990;250:1233–8. doi:10.1126/science.1978757.
3. Kooby DA, Antonesco CR, Brennan MF. Atypical lipomatous tumour/well-differentiated liposarcoma of the extremity and trunk wall: Importance of histological subtype with treatment recommendations. *Ann Surg Oncol*. 2004;11:78–84.
4. Kirova YM, Gambotti L, Rycke YD, Vilcoq JR, Asselain B, Fourquet A, et al. Risk of Second Malignancies After Adjuvant Radiotherapy for Breast Cancer: A Large-Scale, Single-Institution Review. *Int J Radiat Oncol Biol Phys* . 2007;68(2):359–63. doi:10.1016/j.ijrobp.2006.12.011.

5. Gladdy RA, Li-Xuan Q, Moraco N, Edgar MA, Antonescu CR, Alektiar KM, et al. Do Radiation-Associated Soft Tissue Sarcomas Have the Same Prognosis As Sporadic Soft Tissue Sarcomas? *J Clin Oncol* . 2010;28(12):2064–9. doi:10.1200/jco.2009.25.1728.
6. Tuomisto J, Pekkanen J, Kiviranta H, Tukiainen E, Vartiainen T, Viluksela M, et al. Dioxin Cancer Risk — Example of Hormesis? *Dose-Response*. 2005;3(3):332–41. doi:10.2203/dose-response.003.03.004.
7. Tuomisto JT, Pekkanen J, Kiviranta H, Tukiainen E, Vartiainen T, Tuomisto J, et al. Soft-tissue sarcoma and dioxin: A case-control study. *Int J Cancer* . 2004;108(6):893–900. doi:10.1002/ijc.11635.
8. Kleinerman RA, Tucker MA, Tarone RE, Abramson DH, Seddon JM, Stovall M, et al. Risk of New Cancers After Radiotherapy in Long-Term Survivors of Retinoblastoma: An Extended Follow-Up. *J Clin Oncol* . 2005;23(10):2272–9. doi:10.1200/jco.2005.05.054.
9. Singer S. Sabiston textbook of surgery. vol. 1. Philadelphia: Saunders: Elsevier;. p. 768–82.
10. Tan E, Coppola D, Friedman M. Myxofibrosarcoma metastasis to the colon: Case report and review of the literature. *Cancer Treat Commun*. 2016;5:14–6. doi:10.1016/j.ctrc.2015.11.007.
11. Teurneau H, Engellau J, Ghanei I, von Steyern F, Styring E. High Recurrence Rate of Myxofibrosarcoma: The Effect of Radiotherapy Is Not Clear. *Hindawi*. 2019;2019:1–8. doi:10.1155/2019/8517371.

Author biography

Supriya, Post Graduate

Vijay Verma, Post Graduate

Anupam Nanda, Senior Resident

Cite this article: Supriya, Verma V, Nanda A. Myxofibrosarcoma (A rare retroperitoneal tumour)-A case report. *IP Indian J Anat Surg Head, Neck Brain* 2021;7(1):35-38.