BBRC
Bioscience Biotechnology Research Communications

Case Report: A Rare Case Giant Recurrent Retroperitoneal Sarcoma

Tushar Nagtode[1], Rakshit Shah[2], Y. R. Lamture[1], Venkatesh Rewale[1], Ranjit Ambad[3] and Aditya Mundada[1]

[1]*Department Of General Surgery, Jawaharlal Nehru Medical College Wardha, Datta Meghe Institute of Medical Sciences, Wardha, Maharashtra, India*

[2]*Senior Consultant Oncosurgeon, Kailash Cancer Hospital And Research Centre, Vadodara, Gujrat, India*

[3]*Department Of Biochemistry, Datta Meghe Medical College Nagpur. Datta Meghe Institute of Medical Sciences, Wardha, Maharashtra, India*

Corresponding author email: dryrlamture@yahoo.co.in

ABSTRACT

Accounting for only 0.1–0.2 % of total malignancies and grow out of the retroperitoneal organs, retroperitoneal neoplasms are an uncommon yet significant group of neoplasms. Most essential retroperitoneal neoplasms emerge from mesodermal framework with liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma together, representing above 80 % of essential retroperitoneal sarcomas, which is a difficult to treat condition for its localized aggression and clinical particularity. Remaining retroperitoneal masses emerge prevalently from nervous system. Difficulties encountered by surgeons during resection of a retroperitoneal sarcoma of exceptional size, attributable to anatomical site, to nonappearance of an anatomically apparent vascular-lymphatic peduncle and to adhesions contracted with adjoining organs and with large vessels. At present for retroperitoneal soft tissue sarcomas, there's no productive chemotherapy present, and because of radiotoxicity influencing contiguous intra-abdominal organs, radiotherapy has restricted effectiveness yet without precedence in overall survival.

KEY WORDS: NEOPLASM, HIGH GRADE MALIGNANCY.

INTRODUCTION

In current time of surgical practice, large intra-abdominal tumours have become infrequent because of early revelation on regular evaluation. At first, imaging with sonography is liked, however CT scan or MRI scan favoured , when analysis is as yet questionable (Agrawal SP et al., 2015). Identification of tumour causes alarm among patients on account of dread of malignancy prompting psycho-somatic stress disorder. Moreover, huge size of these tumours causes mechanical pressure symptoms on gastrointestinal, respiratory, and urinary tract. Subsequently, a thorough methodology is fundamental to negate secondary impacts alongside treatment of primary tumor (Agrawal SP et al., 2015). Despite advances in treatment, a late finding is principal reason for mortality. Based on age of patient, size and histopathology of tumour , treatment is chosen. In this, we feature an uncommon instance of a postmenopausal lady with history of progressive abdominal swelling over a time of a half year alongside radiological reports expressive of a huge tumour emerging from retro-peritoneum (Agrawal SP et al., 2015).

Case Report: A 50-year-old Indian woman is a housewife, P8L2, with 34 years of marriage, came to oncosurgery outpatient department on 29/04/2019. She complained of gradually increasing size of abdomen since half year along with dull aching pain in swelling since last 2 months. She also complained of increased frequency with sense of incomplete emptying of bladder and had

Biosc Biotech Res Comm P-ISSN: 0974-6455 E-ISSN: 2321-4007

Identifiers and Pagination
Year: 2021 Vol: 14 No (7) Special Issue
Pages: 113-115

Article Information
Received: 18th April 2021
Accepted after revision: 08th June 2021

DOI: http://dx.doi.org/10.21786/bbrc/14.7.27

constipation. She had no history of vomiting or other gastrointestinal problems, colicky pain, or blackouts. Patient underwent laparotomy for abdominal tumour in 2011. No details available. Patient was weighing 40 kg, height 154 cm and BMI 16.8 . On examination patient was afebrile pulse was 68 b/min, blood pressure was 128/64 mm Hg and respiratory rate was 20/min. Her abdomen was distended, abdominal girth was 65 cm at level of umbilicus. Well defined lobulated masses occupying whole abdomen extending till epigastrium with a dull note on percussion present. Her Urine pregnancy test was negative. X ray chest (P-A view) was within normal limits. CA-125 was normal.

Figure 1

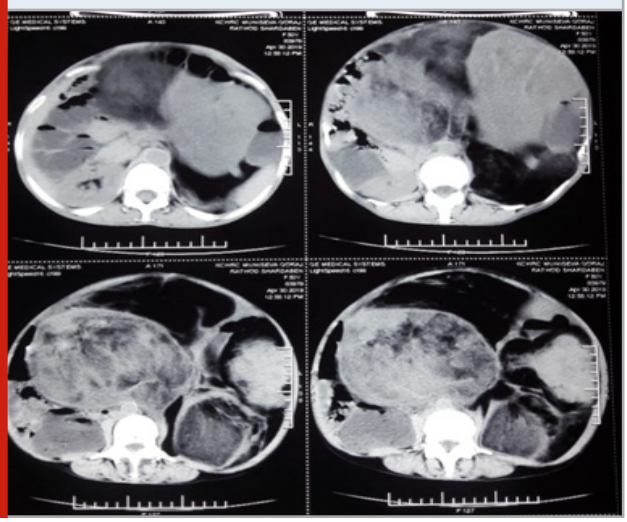
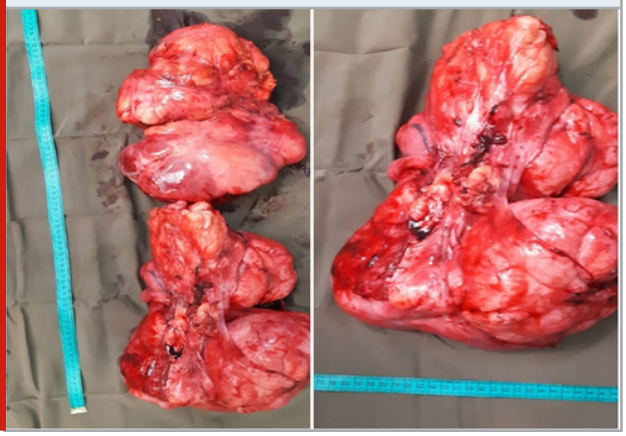


Figure 2



MDCT scan of abdomen and pelvis (plain + contrast) (see Figure 1) was suggestive of multiple lesions, larger lesions measuring approximately 14.9 cm (AP)× 11.6 cm, 12.8 × 9 cm, 10 ×6.6 cm, 8 × 8.7 cm seen arising from the retroperitoneum and occupying most of peritoneal cavity. Exploratory laparotomy –En block Resection of Masses was done on 07/05/2019. Huge three lobulated masses arising from the retroperitoneum seen occupying whole abdomen, was seen displacing bowel loops. Extent of tumour was identified. The tumour mass measured 67 cm × 26 cm and weighed 9.5 kg was removed en bloc (Figure 2). Lymph nodes were not involved. An abdominal drain was placed till post operative day 5. The post-operative period was uneventful. Patient weighed 32kg & abdominal girth was 50 cm at level of umbilicus on post operative day 7. Histopathological examination revealed: Well differentiated liposarcoma.

DISCUSSION

The retroperitoneal liposarcomas are usually huge neoplasms with a low or moderate grade of malignancy. Lungs constitute principle site of distant metastasis (Crago AM et al., 2011). Liposarcoma is the commonest histological type out of all retroperitoneal sarcomas constituting 20-45 %, whereas soft tissue sarcomas constitute 10-15 %. Liposarcomas vary in weight as well as measurement; those more than 20 kg are designated "goliath liposarcomas" and are very uncommon (Fletcher C et al., 2002). We have reported a rare case of a large retroperitoneal liposarcoma very rare, originating from left perinephric fat. CECT abdomen and pelvis is the investigation of choice in patients presumed to be with RPS. CT scan helps to delineate size and extent as well as relationship with nearby vessels, organs, and skeletal structures. It also detects metastasis and gives crucial information regarding plan for surgical resection of tumor (Kumar V et al., 2012).

Liposarcomas are classified based on histology, morphology, and cytogenetic differences. Liposarcomas can be classified into five histologic categories, according to the WHO classification: well differentiated, myxoid, round cell, pleomorphic, and dedifferentiated (Crago AM et al., 2011). Due to common chromosomal translocation, myxoid and round cell types are contemplated as subtypes (Hashimoto Y et al., 2010). Undifferentiated and pleomorphic are highly malignant, aggressive and can metastasize, whereas well differentiated and myxoid/round cell tumours are low grade and have good prognosis. Well-differentiated and undifferentiated tumours constitute well established biological group (Crago AM et al., 2011). Well differentiated liposarcomas are locally aggressive but have low metastatic potential and identified as well encapsulated, homogeneous lesion isodense with adipose tissue with thickened septa. However undifferentiated liposarcomas emerge from histological aberration in relation to well differentiated ones. Further loss of differentiation decides cellular transformation. Its distinctive of recurrence of disease, it takes place in 20% at 1st recurrence and in 44% at 2nd (Lewis JJ et al., 1998).

The histological type of sarcoma cannot be predicted with certainty by imaging. The presence of fat in the tumor on a CT scan can sometimes be used to diagnose liposarcoma. Unfortunately, imaging cannot predict the tumor grade, which has prognostic value (Kransdorf MJ 2002). Resection of a retroperitoneal sarcoma of exceptional size, attributable to anatomical site, to nonappearance of an anatomically apparent vascular-lymphatic peduncle that makes it hard to obtain safe

margin and to adhesions contracted with adjoining organs and with large vessels. It is more challenging in case of recurrence, due to distorted anatomy by previous surgery (Caizzone A 2015). Liposarcomas, both well-differentiated and dedifferentiated, are difficult to treat. Surgical excision of a localized tumor is the cornerstone of treatment. Patients with advanced tumors do, in fact, have few therapy alternatives. Anthracycline-based regimens have a poor response rate and a short median PFS. Nonetheless, trabectedin and eribulin have recently been approved for advanced liposarcoma treatment. Furthermore, the FDA has approved olaratumab (in conjunction with doxorubicin) (McGovern Y et al., 2017).

The outcome of patients with WDLS and DDLS is determined by the extent of surgical resection, as well as the tumor's location and histologic subtype. Patients with dedifferentiated histology or the retroperitoneal area have a higher risk of recurrence. We now know that individuals with rapidly developing or partially resectable tumors have poor post-surgery outcomes, hence these patients should be treated non-operatively. In WDLS and DDLS, low response to radio and chemotherapy is observed, despite the fact that medicines targeting chromosome 12 gene products, MDM2 and CDK4, have shown promise in preclinical investigations and are currently being studied in clinical trials (Crago AM 2011).

Several interesting medicines are currently being evaluated in advanced WDLS and DDLS, including a stage 2/3 DDLS evaluation of selinexor. CDK4 and MDM2 inhibitors are showing promise, particularly as prospective combo therapies with traditional chemotherapy. Immunotherapy using checkpoint inhibition is a rapidly growing area within the story of systemic treatment for liposarcoma, and early results are promising, particularly in the case of undifferentiated pleomorphic sarcoma but also dedifferentiated liposarcoma (McGovern Y et al., 2017). In medical literature, many studies on different types of sarcoma10-13 and novel therapeutic aspects14 have been reported. Extra targets, such as ZIC1, TOP2A, AURKA, and IGF-1R, have been discovered by cell line, tissue microarray, and genomic studies, which could form the basis of future treatment (Crago AM 2011).

CONCLUSION

The whole surgical resection is presently best treatment for liposarcoma. Most of sufferers with RPS, in due course have recurrence and get perished of disease. It was hoping that a multimodality approach, which includes whole surgical resection and targeted therapies, will drastically enhance affected person outcomes.

REFERENCES

Agrawal SP, Rath SK, Aher GS, Gavali UG (2015). Large Ovarian Tumor: A Case Report;3(3):3.

Caizzone A, Saladino E, Fleres F, Paviglianiti C, Iaropoli F, Mazzeo C, et al (2015). Giant retroperitoneal liposarcoma: Case report and review of the literature. Int J Surg Case Rep;9:23–6.

Chaudhary, Minal, Kanhaiya Bajaj, Dipak Ghatage, Shruti Bohra, and Nitin Bhola ((February 2015)). Lesion of Dual Nature - Carcinoma or Sarcoma: A Histopathologic Dilemma." Clinical Cancer Investigation Journal 4, no. 1: 43–46. https://doi.org/10.4103/2278-0513.149038.

Crago AM, Singer S (2011 Jul). Clinical and molecular approaches to well differentiated and dedifferentiated liposarcoma. Curr Opin Oncol;23(4):373–8.

Fletcher C (2002). Pathology and genetics of tumors of soft tissue and bone. World Health Organ Classif Tumors;4:35–46.

Gupte, Yash, Parth Godhiwala, Amrutha Garikapati, Sunil Kumar, Sourya Acharya, Samarth Shukla, and Chetan Rathi (December 2020). Right Atrial Sarcoma Masquerading as Myxoma Presenting with Fatal Cardiac Tamponade: A Rare Case Report." Medical Science 24, no. 106: 4374–78.

Hashimoto Y, Hatakeyama S, Tachiwada T, Yoneyama T, Koie T, Kamimura N, et al (2010). Surgical treatment of a giant liposarcoma in a Japanese man. Adv Urol;2010:943073.

Khatib, Mahalaqua Nazli, Abhay Gaidhane, Shilpa Gaidhane, and Zahiruddin Quazi Syed ((2018)). Ghrelin as a Promising Therapeutic Option for Cancer Cachexia." Cellular Physiology And Biochemistry 48, no. 5: 2172–88. https://doi.org/10.1159/000492559.

Kransdorf MJ, Bancroft LW, Peterson JJ, Murphey MD, Foster WC, Temple HT (2002 Jul). Imaging of Fatty Tumors: Distinction of Lipoma and Well-differentiated Liposarcoma. Radiology; 1;224(1):99–104.

Kumar V, Misra S, Chaturvedi A (2012 Sep). Retroperitoneal Sarcomas- A Challenging Problem. Indian J Surg Oncol;3(3):215–21.

Lewis JJ, Leung D, Woodruff JM, Brennan MF (1998 Sep). Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution. Ann Surg;228(3):355–65.

McGovern Y, Zhou CD, Jones RL (2017). Systemic Therapy in Metastatic or Unresectable Well-Differentiated/ Dedifferentiated Liposarcoma. Front Oncol;7:292.

Nimonkar, Pranali, Nitin Bhola, Anendd Jadhav, Anuj Jain, Rajiv Borle, Rajul Ranka, and Minal Chaudhary ((2016)). Myxoid Chondrosarcoma of Maxilla in a Pediatric Patient: A Rare Case Report." Case Reports In Oncological Medicine 2016. https://doi.org/10.1155/2016/5419737.

Yeola, Meenakshi, Kushagra Singh, Aditya Mehta, and Roshani S. Manekk (November 2, 2020). "Atypical Presentation of Recurrent High-Grade Myxofibrosarcoma - A Case Report." Journal of Evolution Of Medical And Dental Sciences-Jemds 9, no. 44: 3348–50. https://doi.org/10.14260/jemds/2020/736.