Giant Retroperitoneal Sarcoma

Nimesh Shah, Jignesh Gandhi, Bhagvat Vikrant M.

Abstract

Retroperitoneal sarcoma is a rare malignancy often insidious in onset. At initial presentation, abdominal mass, pain or obstructive symptoms are the most common complaints. We report a case of retroperitoneal sarcoma in a 32 year female presenting with fullness in the abdomen since one year.

Introduction

Soft-tissue sarcomas are relatively rare with approximately 8,600 new cases annually and represent less than 1% of all newly diagnosed malignancies. Retroperitoneal sarcomas are malignant tumours arising from mesenchymal cells, which are usually located in muscle, fat, and connective tissues. One-third of malignant tumours located in the retroperitoneum are sarcomas, and approximately 15% of soft tissue sarcomas arise in the retroperitoneum.[1] According to the World Health Organisation (WHO), soft-tissue liposarcomas are categorised into five distinct histological subtypes: well-differentiated, dedifferentiated, myxoid, pleomorphic and mixed type. Retroperitoneal sarcomas have varying clinical courses depending on their histological subtype and grade.[1,2]

The typical presenting symptom is an abdominal mass, pain, or symptoms related to compression of adjacent abdominal structures. Review of the literature reveals that the most common ages of presentation are in the 5th or 6th decade, with similar distribution among the sexes. Presentation is often complicated by metastasis or involvement of local structures.

Dept. of General Surgery, Global Hospital, Opposite Shirodkar High School,Parel, Mumbai.

Case Report

A 32 year-old female was admitted with distension of abdomen since one year. The patient had a past history of open hysterectomy done seven years back. Abdominal examination showed distension all over the abdomen (Fig. 1). Her blood

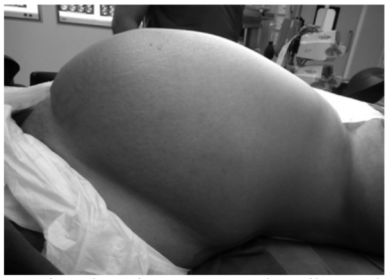

Fig. 1: Showing the preoperative distension of abdomen

investigations were within normal range. The computed tomography done revealed a large retroperitoneal liposarcoma with displacement of the bowel loops and pancreas to the left side, right kidney antero-inferiorly and liver superiorly (Fig. 2). The

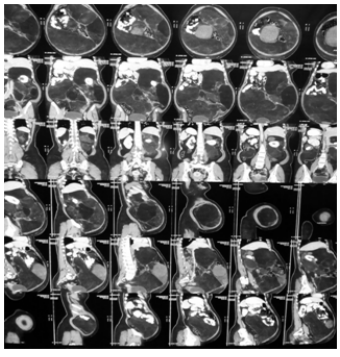

Fig. 2: CT scan showing the size and extent of the tumour

right ureter was seen coursing through the mass lesion. Exploratory laparotomy was performed and the liposarcoma extended superiorly upto the liver and suprarenal and inferiorly upto the pelvis. The colon and terminal ileum was adherent to the liposarcoma. The liposarcoma was crossing the midline and going to the sigmoid and the duodenojejunal flexure. The liposarcoma was encasing the right kidney and the ureter (Fig. 3). Total

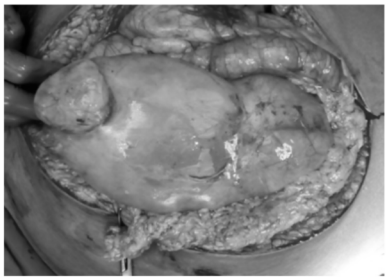

Fig. 3: Showing the intraoperative extent of the tumour

enmass excision of the liposarcoma (Fig. 4) was

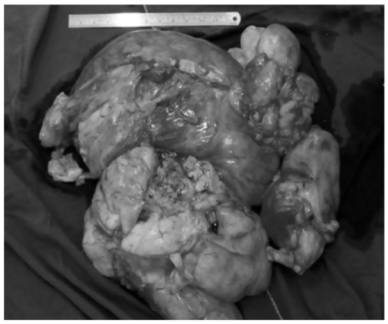

Fig. 4: Showing the size of the excised tumour

carried out. The patient had a uneventful post operative course and discharged on the 12th day. The histopathological report confirmed the diagnosis of retroperitoneal liposarcoma. The patient would be later subjected to postoperative chemotherapy and radiotherapy.

Discussion

Retroperitoneal tumours are an extremely heterogeneous group of neoplasms, 85% of which are malignant. Liposarcomas constitute between 45-55% of retroperitoneal masses.[4] Age at presentation is younger compared with most other malignancies, with many being diagnosed.

Between 54-65 years of age.[5] There is an equal male/female ratio.[1] The distribution of soft tissue sarcomas by anatomic site can be found in an article by Lawrence et al.[6]

Retroperitoneal sarcomas present 80% of the time as an asymptomatic abdominal mass. Symptoms can also be related to mass effect or local invasion which may lead to pain, gastrointestinal obstruction, feelings of early satiety, and weight loss. In addition, neurologic and muscular skeletal symptoms are referred to the lower extremities.[7]

Histopathologic variety is the main prognostic factor. Five histologic types are recognised. Well differentiated liposarcoma represents around 30% like our case and has the best prognosis. The myxoid type is the most frequent liposarcoma, constituting around 50% of all tumours. It has a less favourable progression, as it often recurs early. The pleomorphic, round cell and undifferentiated types display the worst prognosis.[4]

After a physical examination CT scan provides an excellent understanding of the relationship between nearby structures and is critical to preoperative planning. A patient presenting with a palpable abdominal mass, should have a high-resolution, thin-cut CT scan with intravenous and oral contrast since these images allow for further distinction between intra-abdominal and retroperitoneal structures. This allows a discussion of the need for biopsy if indicated, the operative plan, and the

preparedness of the operative team, as well as a discussion with the patient regarding the risks and benefits. The differential diagnosis includes a primary neoplasm arising from a retroperitoneal visceral structure (e.g., pancreas, adrenal glands, kidneys, and duodenum), a retroperitoneal sarcoma, a lymphoma, or a metastatic lesion.[5]

The optimal treatment for patients with localised, resectable retroperitoneal sarcomas is surgery with gross and microscopically negative margins. Complete surgical resection frequently requires en-bloc resection of adjacent viscera.[8] The kidney was the most frequently resected organ (36%) followed by segmental resection of the large bowel, spleen, and pancreas.[9]

The addition of adjuvant radiation therapy to surgical resection is associated with a reduced risk of local recurrence and a longer recurrence-free interval. However, it does not improve overall survival. Studies have demonstrated the advantages of preoperative radiotherapy in the management of marginally resectable retroperitoneal sarcomas. The benefits of pre-operative radiation are multiple.[3,10] It allows for the gross tumour volume to be readily definable for accurate treatment planning. Moreover, the tumour displaces radiosensitive viscera. Thus, no adhesions and tethering of bowel to the tumour bed can occur and the tumour is treated in situ.

Another treatment modality is intra-operative radiotherapy (IORT) which is targeted to a specific region allowing for maximum doses of radiation to the tumour bed. Studies show that IORT improves tumour control in the field. However, it does not influence recurrence-free or overall survival rates.[9,10]

Conclusion

The review of the literature emphasises that the management of retroperitoneal sarcomas consists of complete resection of the tumour followed by adjuvant radiotherapy which reduces local recurrence but does not affect overall survival and combined with surveillance for early detection of recurrence or metastases. Imaging studies are essential for proper preoperative planning and allow assessment of respectability prior surgery; preoperative radiotherapy can be considered in patients with questionably resectable tumours. The patient should be closely followed with regular physical examinations and imaging studies such as chest X-rays and computed tomography scans.

References

1. Windham TC, Pisters PW. Retroperitoneal sarcomas. *Cancer Control* 2005;12(1):36-43. [Pubmed]
2. Kawano R, Nishie A, Yoshimitsu K, et al. Retroperitoneal well-differentiated inflammatory liposarcoma: a diagnostic dilemma. *Radiat Med* 2008;26(7):450-3
3. Stoeckle E, Coindre JM, Bonvalot S, et al. Prognostic factors in retroperitoneal sarcoma: a multivariate analysis of a series of 165 patients of the French Cancer Center Federation Sarcoma Group. *Cancer* 2001;92(2):359-68. [CrossRef] [Pubmed]
4. Leão P, Vilaça S, Oliveira M, Falcão J. Giant recurrent retroperitoneal liposacrcoma initially presenting as inguinal hernia: review of literature. *Int Journal of Surgery case Report* 2012;3(3):103-6. [CrossRef] [Pubmed]

5. John E Mullinax, Jonathan S Zager, Ricardo J Gonzalez. Current Diagnosis and Management of Retroperitoneal Sarcoma. *Cancer Control* 2011;18(3):177-87. [Pubmed]
6. Lawrence W Jr, Donegan WL, Natarajan N, et al. Adult soft tissue sarcomas. A pattern of care survey of the American College of Surgeons. *Ann Surg* 1987;205(4):349-59. [CrossRef] [Pubmed]
7. Wanchick K, Lucha P. Dedifferentiated retroperitoneal liposarcoma presenting as lower gastrointestinal bleeding, a case report and review of the literature. *Military Medicine* 2009; 174(3):328-30. [Pubmed]
8. Pisters PW, O'Sullivan B. Retroperitoneal sarcoma: combined modality treatment approaches. *Curr Opin Oncol* 2002;14(4):400-5. [CrossRef] [Pubmed]
9. Hassan I, Park SZ, Donohue JH, et al. Operative management of primary retroperitoneal sarcomas: a reappraisal of an institutional experience. *Annals of Surgery* 2004;239(2):244-50. [CrossRef] [Pubmed]
10. Mendenhall WM, Zlotecki RA, Hochwald SN, et al. Retroperitoneal soft tissue sarcoma. Cancer 2005;104(4):669-75. [CrossRef] [Pubmed

Drugs for rheumatoid arthritis: looking backwards to move forwards

New combinations of older drugs can be effective and affordable

These steps include the introduction of methotrexate in 1980 and the first inhibitors of tumour necrosis factor (TNF) in 2000.

In parallel, trials have also continued to test old drugs used in new more modern ways, starting with the combination of methotrexate, sulphasalazine, and hydroxychloroquine. The real surprise came from a recent demonstration that this combination was as effective as the combination of methotrexate and the TNF inhibitor etanercept.

The most common TNF inhibitor was adalimumab (58/101) and the most common drug combination was methotrexate and leflunomide (62/104).

There were also differences. At six months, TNF inhibitors were associated with lower disease activity relative to drug combinations, a difference not seen at 12 months, and indicating the rapid effect of TNF inhibition.

The good news is that a combination of traditional drugs was not inferior to a biological option as found in previous trials.

Another limitation is that rheumatoid arthritis is a chronic disease. When active treatment stops, active disease often restarts. One option, at least in responders, is to continue treatment, but at a reduced dose.

If we were to take the same approach to rheumatoid arthritis then we would combine drugs at the start of treatment and not wait for traditional monotherapies to fail.

Considering rheumatoid arthritis care today, the best way to improve overall outcomes is to act earlier.

Pierre Miossec, BMJ, 2015, Vol 350, 8