A Case Of Retroperitoneal Myxofibrosarcoma

V Abhishek, M Ajitha, U Mohan, B Shivswamy

Citation

V Abhishek, M Ajitha, U Mohan, B Shivswamy. *A Case Of Retroperitoneal Myxofibrosarcoma*. The Internet Journal of Surgery. 2012 Volume 28 Number 3.

Abstract

A 56-year-old male visited a local physician with intermittent abdominal pain. Ultrasonography revealed a mass of about 6cm arising from bowel and the patient was referred to our hospital. Clinically, a freely mobile mass was palpable in the right para-umbilical region. Ultrasound-guided fine-needle aspiration cytology (FNAC) showed features of a spindle cell tumor. On abdominal computed tomography, a tumor measuring 7x10x6cm in the right anterior para-renal space with uneven contrast-enhanced effects was recognized displacing duodenum and inferior vena cava and encasing the aorta at L2 level. A diagnosis of retroperitoneal tumor was made, and surgery was performed. Histopathological examination revealed myxoid areas with pleomorphic stellate and spindle cells. The nucleoli were pleomorphic and hyperchromatic with prominent nucleoli. Many branching vessels were seen throughout with bizarre multinucleate giant cells. Features were consistent with high-grade myxofibrosarcoma.

Retroperitoneal sarcomas represent 0.1% of all malignancies. Population estimates place the yearly incidence at 2.7 cases per million people[1]. The most common histology is liposarcoma, followed by leiomyosarcoma. Other rare subtypes include ?brosarcoma and malignant peripheral nerve sheath tumors. First described in 1977, myxofibrosarcoma is one of the rare sarcoma subtypes affecting mainly the extremities and skin. The largest study of 75 cases of myxofibrosarcoma was taken up by Mentzel et al. with only 2 of those in the retroperitoneum[2]. Another case report of a retroperitoneal myxofibrosarcoma is by Takeuchi et al. of Ohashi Hospital Japan, making retroperitoneal myxofibrosarcoma one of the rare tumors.

CASE REPORT

A 56-year-old male presented with history of intermittent abdominal pain since 4 months for which he consulted a local physician. Ultrasonography revealed a mass of about 6cm arising from bowel and the patient was referred to our hospital. There was no history of weight loss or altered bowel habits and all routine blood investigations were unremarkable. Clinically, a freely mobile mass was palpable in the right para-umbilical region which did not move with respiration. On abdominal computed tomography, a tumor measuring 7x10x6cm in the right anterior para-renal space with uneven contrast-enhanced effects was recognized displacing duodenum and inferior vena cava and encasing the aorta at L2 level (Fig. 1, 2). CT also revealed prostatic median lobe hypertrophy and bilateral renal simple cysts Ultrasound-guided fine-needle aspiration cytology (FNAC) showed features of a spindle cell tumor.

A diagnosis of a retroperitoneal tumor was made, and surgery was performed.

Histopathological examination: Cut section showed grey-white firm and soft yellow areas with vague nodular pattern at places. Microscopy showed myxoid areas with pleomorphic stellate and spindle cells. The nucleoli were pleomorphic and hyperchromatic with prominent nucleoli. Many branching vessels were seen throughout with bizarre multinucleate giant cells (Fig. 5, 6) Features were consistent with high-grade myxofibrosarcoma.

There were no additional complications after the operation and the patient was discharged after one week of operation.

Figure 1

Fig. 1: CT picture of retroperitoneal tumor

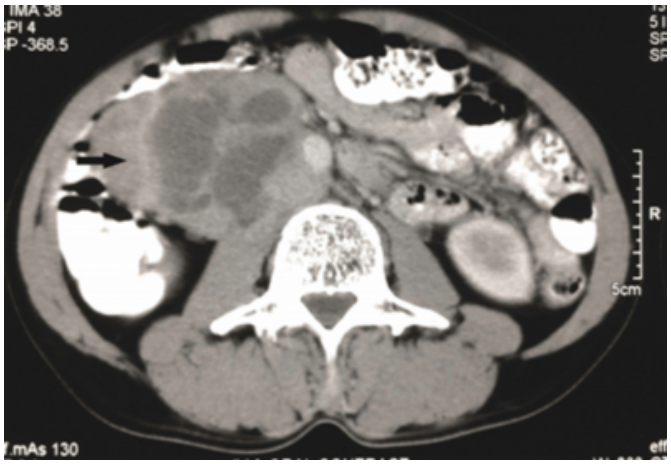


Figure 2

Fig. 2: CT picture of retroperitoneal tumor

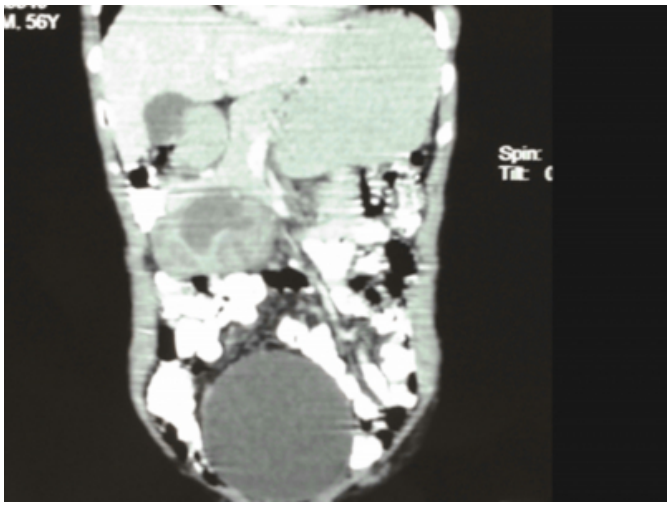


Figure 3

Fig. 3: Intra-operative finding: Tumor encasing the inferior vena cava

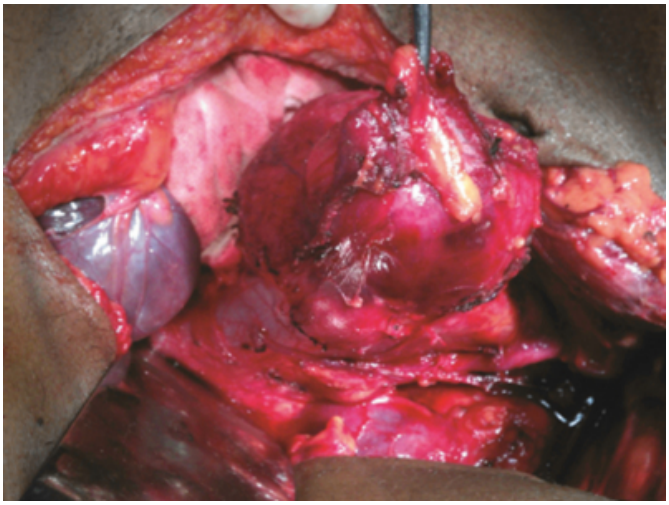


Figure 4

Fig. 4: Specimen of retroperitoneal tumor

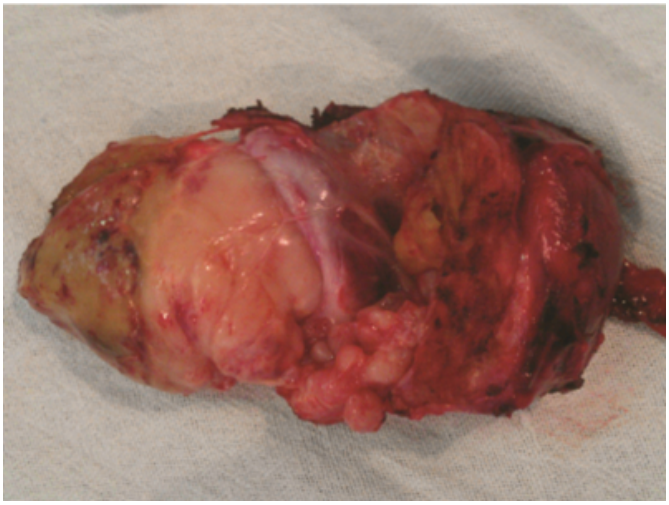


Figure 5

Fig. 5: Histopathology showing myxoid areas with pleomorphic stellate and spindle cells (hematoxylin and eosin, 15x)

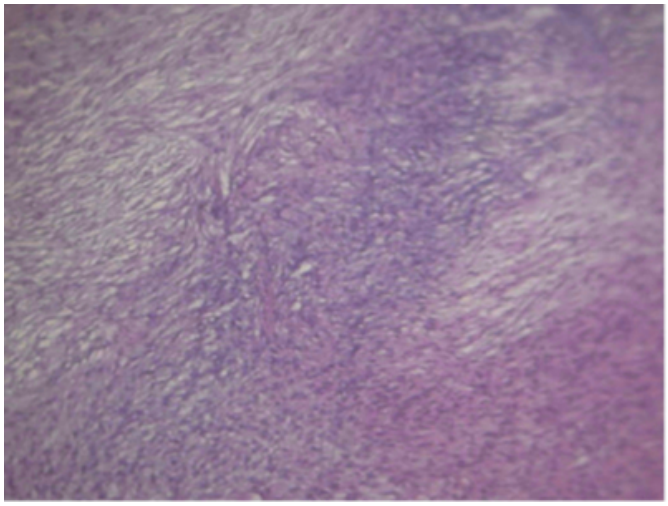
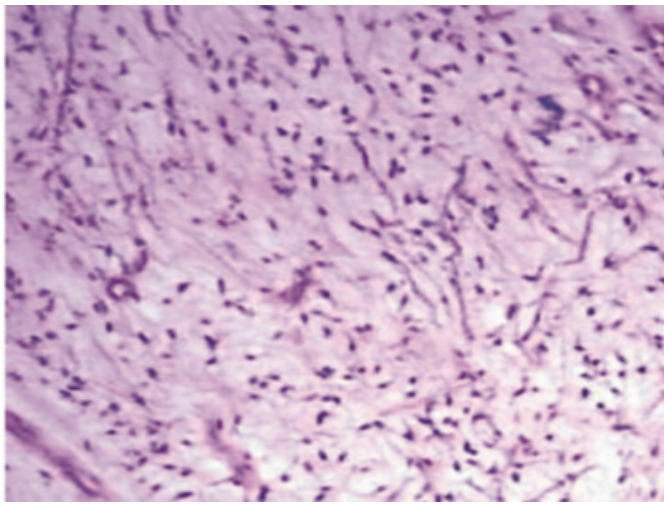


Figure 6

Fig. 6: Histopathology showing branching vessels with bizarre multinucleate giant cells (hematoxylin and eosin, 75x)



DISCUSSION

Retroperitoneal sarcomas are rare tumors with the most common histology being liposarcoma (61%), followed by leiomyosarcoma (30%)[3] and other rare varieties like fibrosarcoma and peripheral nerve sheath tumors.

Retroperitoneal tumors typically have vague presenting symptoms. These tumors are provided with a well-concealed, widely expansible area, leading to the development of large masses with local and distant metastases before the patient becomes symptomatic. Todd et al. reported that the most common clinical picture of retroperitoneal sarcoma cases at presentation includes back pain and weight loss (37.5% of patients with either symptom), with fatigue (25%), increased abdominal girth (12.5%), and fever or night sweats (12.5%) also noted.[4]

These patients exhibited symptoms for an average 3.5 months before presentation. Twenty-five percent of patients have masses discovered incidentally during routine examination or abdominal surgery (cholecystectomy). The studies of Cody et al. and Braasch et al. both reported abdominal pain and weight loss were the most common symptoms[5,6].

The diagnosis of retroperitoneal tumor is aided by imaging studies which contribute greatly to delineating the size, location, and character of a mass. Todd et al. pointed out that CT was the preferred method for evaluation of a known mass prior to exploratory laparotomy.[4] A myxofibrosarcoma on CT and magnetic resonance imaging typically has uneven contrast-enhanced effects with both solid and cystic components.

The myxoid variant of malignant fibrous histiocytoma, introduced by Weiss and Enzinger in 1977, is not of true histiocytic origin and, increasingly, the term histiocytoma seems unjustified in the light of modern histopathology studies[7]. The Scandinavian authors, who have contributed most of the limited work published to date, instead used the term myxofibrosarcoma, which highlights the myxoid matrix and implies fibroblastic origin.

Myxofibrosarcoma, also often known as the myxoid variant of MFH, is one of the most common sarcomas in the extremities of elderly people and occurs more frequently in dermal and subcutaneous tissues than in deeper tissues[7]. Deeper structure locations are very rare and tend to be more aggressive. Atypical locations are thorax, retroperitoneum and pelvis[2]. Grossly, the tumor usually consists of multiple gelatinous nodules composed histologically of noncohesive, mostly fusiform but also round or stellate tumor cells. Characteristic morphologic features important in diagnosis are a prominent vascular pattern of curvilinear small capillaries with perivascular tumor cell accentuation and a striking inflammatory infiltrate[9].

The tendency of myxofibrosarcoma to be of progressively higher grade in recurrences by becoming gradually more cellular, more pleomorphic, more mitotically active, and more necrotic seems to be a distinctive feature and is in contrast to most other sarcomas.

DIAGNOSTIC CRITERIA

IMMUNOHISTOLOGY

Figure 7

Vimentin	Positive
Smooth muscle actin	occasionally focally weak positive
S100	Negative
CD34	occasionally focally weak positive
MUC4	Negative

LOW GRADE

INTERMEDIATE GRADE

HIGH GRADE

Surgical resection remains the mainstay of treatment for retroperitoneal sarcoma with resection rates ranging from 54-88% in several case series[9]. Complete surgical resection with at least 3cm margins is the treatment of choice but is rarely feasible due to invasion of adjacent structures by the tumor. Erzen et al. pointed out that some tumors might need to be resected en bloc with adjacent structures in order to obtain free margins[11]. In that report, at least one organ was resected completely or partially in 118 patients. The most frequent organ resected was the bowel. The feasibility of complete resection is one of the important factors influencing the survival of patients.

Curative surgery is difficult with large retroperitoneal sarcomas and those in close proximity to vital structures and involving adjacent organs. The rate of complete resection has varied in the literature from 38% to 95%[12]. Liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma represented 66% of the tumors in a review of 165 cases of retroperitoneal sarcoma by Stoeckle et al. Complete excision was achieved in 94 of 145 patients who had no metastasis. Aggressive surgery remains mandatory in retroperitoneal sarcoma. Despite these efforts, local recurrence is common. The actuarial overall 5-year rate was 46%[12]. Prognosis is largely related to the attainment of clear surgical margins[13] and to the degree of differentiation of the sarcoma at the time of presentation. Multivariate analysis of the cases showed that a higher grade malignancy, large tumors, and deep tumors were correlated significantly with decreased metastasis-free survival. Inadequate local treatment was correlated with local recurrence, and a high grade of malignancy was correlated with decreased overall survival[14].

With the exception of childhood soft-tissue sarcomas such as rhabdomyosarcoma and extraskeletal Ewing's sarcoma, the benefits of adjuvant chemotherapy for soft-tissue sarcoma remain controversial. A study of a series of 183 patients with truncal or retroperitoneal sarcomas by Singer et al. suggests a poor outcome (3 to 4.6-fold increase in the risk of death) for patients who received either preoperative or postoperative chemotherapy[15].

In a retrospective review of 23 patients with retroperitoneal sarcomas by Bautista et al., liposarcomas were the most common pathology (61%) and leiomyosarcomas were second (30%). Low-grade sarcomas overall accounted for 62 per cent of the total group. Low-grade tumors independent of the histologic type exhibited good prognosis for long-term survival with a median survival of 44 months. In contrast, intermediate- or high-grade tumors were associated with a median survival of only 9 months ($P < 0.02$). Tumor grade was identified as an important prognostic factor with low-grade retroperitoneal sarcomas being associated with significantly longer median survival times. Liposarcomas were associated with a median survival time of 33 months and leiomyosarcomas with 24 months.

The histologic tumor type, independent of grade, did not show a significant survival difference. In many other studies, the histologic type, independent of tumor grade, was not shown to be important for prognostic evaluation.

In the study by Stoeckle et al., 84% of the tumors (165 cases) were high or intermediate grade. There was no significant survival difference between different histologic type tumors. A high grade affected local recurrence, metastatic recurrence, and survival.

References

1. Porter GA, Baxter NN, Pisters PW: Retroperitoneal sarcoma: a population-based analysis of epidemiology, surgery, and radiotherapy. Cancer; 2006; 106(7):1610-1616
2. Mentzel T, Calonje E, Wadden C, Camplejohn RS, Beham A, Smith MA, Fletcher CD: Myxofibrosarcoma. Clinicopathologic analysis of 75 cases with emphasis on the low-grade variant. Am J Surg Pathol; 1996; 20(4): 391-405.
3. Bautista N, Su W, O'Connell TX: Retroperitoneal soft tissue sarcomas: prognosis and treatment of primary and recurrent disease. Am Surg; 2000; 66: 832-6.
4. Todd CS, Michael H, and Sutton G: Retroperitoneal leiomyosarcoma: eight cases and a literature review. Gynecol Oncol; 1995; 59: 333-7.
5. Cody HS, Turnball AD, Fortner JG, Hajdu SI: The continuing challenge of retroperitoneal sarcomas. Cancer; 1981; 47: 2147-52.
6. Braasch JM and Mon AB: Primary retroperitoneal tumors. Surg Clin North Am; 1967; 47: 663-77.
7. Weiss SW, Enzinger FM: Myxoid variant of malignant fibrous histiocytoma. Cancer; 1977; 39: 1672-85.
8. Merck C, Angervall L, Kindblom LG, Oden A: Myxofibrosarcoma. A malignant soft tissue tumor of

fibroblastic histiocytic origin. A clinicopathologic and prognostic study of 110 cases using a multivariate analysis. APMIS; 1983; 91(Suppl 282): 1-40.
9. Angervall L, Kindblom LG, Merck C: Myxofibrosarcoma. A study of 30 cases. Acta Pathol Microbiol Scand (A); 1977; 85: 127-40.
10. Stoeckle E, Coindre JM, Bonvalot S, Kantor G, Terrier P, Bonichon F, Nguyen Bui B: Prognostic factors in retroperitoneal sarcoma: a multivariate analysis of a series of 165 patients of the French Cancer Center Federation SarcomaGroup. Cancer; 2001; 92: 359-68.
11. Erzen D, Sencar M, and Novak J: Retroperitoneal sarcoma: 25 years of experience with aggressive surgical treatment at the institute of oncology, Ljubljana. J Surg Oncol; 2005; 91: 1-9.
12. Catton CN, O'Sullivan B, Kotwall C, Cummings B, Hao Y,Fornasier V: Outcome and prognosis in retroperitoneal softtissue sarcoma. Int J Radiat Oncol Biol Phys; 1994; 29: 1005-10.
13. McGrath P, Neifeld JP, Lawrence W, DeMay RM, Kay S,Horsley JS 3rd, Parker GA. Improved survival following complete excision of retroperitoneal sarcomas. Ann Surg; 1984; 200: 200-4.
14. Svarvar C, Bohling T, Berlin O, Gustafson P, Folleras G, Bjerkehagen B, Domanski HA, Sundby Hall K, Tukiainen E,Blomqvist C: Clinical course of nonvisceral soft tissue leiomyosarcoma in 225 patients from the Scandinavian Sarcoma Group. Cancer; 2007; 109: 282-91.
15. Singer S, Cirsib JM, Demetri GD, Healey EA, Marcus K, Eberiein TJ: Prognostic factors predictive of survival for truncal and retroperitoneal soft-tissue sarcomas. Ann Surg; 1995; 221: 185-95.

Author Information

V Abhishek, M S General Surgery
Postgraduate, Department Of General Surgery, Victoria Hospital, Bangalore Medical College And Research Institute

M B Ajitha, M S General Surgery
Department Of General Surgery, Victoria Hospital, Bangalore Medical College And Research Institute

Ullikashi Mohan, M S
Department Of General Surgery, Victoria Hospital, Bangalore Medical College And Research Institute

B S Shivswamy, M S
Department Of General Surgery, Victoria Hospital, Bangalore Medical College And Research Institute