Case Report

Retroperitoneal leiomyosarcoma in a young adult presenting as a huge abdominal mass

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ABSTRACT

Retroperitoneal sarcomas are rare tumors accounting for only 1–2% of all solid malignancies. Only 10–20% of sarcomas are retroperitoneal sarcomas, and the overall incidence is 0.3–0.4%/100,000 of the population. The peak incidence is in the fifth decade of life. These tumors grow very large as they have a very large retroperitoneal space to grow and producing very vague and mild symptoms like abdominal fullness and pain. We are hereby presenting a case of a 24-year-old female who presented with increasing abdominal girth, intermittent abdominal discomfort, palpable abdominal mass, and a 5 kg weight loss.

Key words: Leiomyosarcoma, retroperitoneum, soft-tissue tumors

INTRODUCTION

Retroperitoneal sarcomas are rare tumors accounting for only 1–2% of all solid malignancies. Of all sarcomas, the majority occur outside of the retroperitoneum. Only 10–20% of sarcomas are retroperitoneal sarcomas, and the overall incidence is 0.3–0.4%/100,000 of the population.[1] The peak incidence is in the fifth decade of life, although they can occur in any age group. Patients with sarcomas present late, because these tumors arise in the large potential spaces of the retroperitoneum and can grow very large without producing symptoms.[2,3] Moreover, when symptoms do occur, they are nonspecific, such as abdominal pain and fullness, and are easily dismissed as being caused by other less serious processes.[4] Retroperitoneal sarcomas, therefore, are usually very large at the time of presentation, and there is a need to diagnose these rare tumors at the earliest so as to begin early treatment.

Access this article online	
Quick Response Code:	Website: www.ccij-online.org
	DOI: 10.4103/2278-0513.154276

CASE REPORT

A 24-year-old female was in good health, when she was admitted in our hospital with a 2-month history of increasing abdominal girth, intermittent abdominal discomfort, palpable abdominal mass, and a 5 kg weight loss. She also complained of anorexia and increasing fatigue. The menstrual cycles were normal, and there was no history of fever. Physical examination revealed a 13 cm palpable mass over the left middle portion of the abdomen. X-ray of kidney, ureters and bladder revealed a displaced transverse colon. Ultrasonographic study revealed a heterogeneous mass in the abdomen which displaced the surrounding structures. Computed tomography (CT) of the abdomen also revealed an approximately 12 cm × 10 cm × 8 cm heterogeneous, retroperitoneal mass with contrast enhancement, which did not involve any major organ or vessels. Laboratory investigations revealed hemoglobin of 7.5 g/dl (normal range: 11–16 g/dl) with a total leukocyte count of 5200/μL (4000–11,000/μL) and platelet count of 165,000/dl (150,000–400,000/dl) blood urea/creatine and liver function tests were normal. She underwent an exploratory laparotomy with resection of a 430 g, irregular; bosselated and firm retroperitoneal mass [Figure 1]. The tumor was not attached to the duodenum and other great vessels. Pathological examination revealed a tumor tissue comprising of spindle cells arranged in nodules and fascicles [Figure 2]. Smooth muscle cell differentiation was

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seen in many areas. Cells have elongated blunt-ended nuclei with cytoplasmic vacuoles indenting the nuclei [Figure 3]. Few areas showed nuclear pleomorphism and large bizzare nuclei, with areas of necrosis and calcifications. Sections also show 10 mitoses/10 high power fields. Surgical margins were free from tumor. Smooth muscle actin shows strong positivity [Figure 4]. c-kit and CD34 were negative. The features are suggestive of a low-grade malignant tumor showing myogenous differentiation consistent with leiomyosarcoma. Patient was referred to a higher oncosurgical center for further decision/treatment and follow-up.

DISCUSSION

Soft-tissue sarcomas are rare tumors accounting for approximately 1% of all adult malignancies. Fifty percent of these occur in the extremities followed by retroperitoneum and trunk.[5] Retroperitoneal soft-tissue sarcomas account for 13% of all adult soft-tissue sarcomas with malignant fibrous histiocytomas and liposarcomas being the most common.

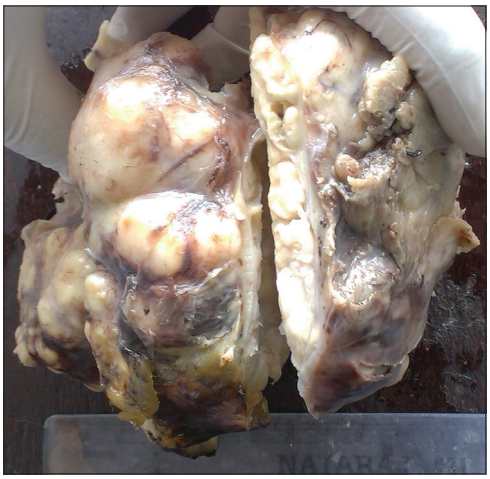

Figure 1: Gross image showing irregular, bosselated and firm soft-tissue tumor

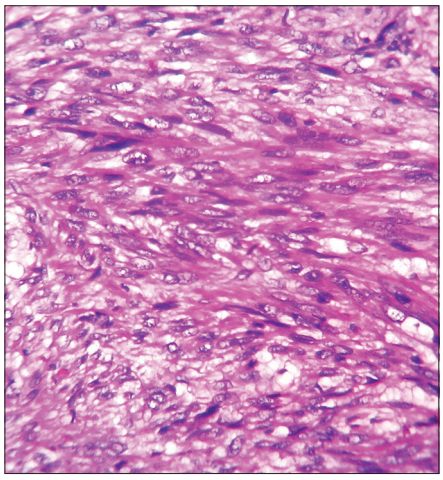

Figure 3: Micrograph showing elongated blunt ended, pleomorphic nuclei with cytoplasmic vacuoles (H and E, ×40)

Retroperitoneal leiomyosarcoma occurs most commonly in the fifth to seventh decade.[6] 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*.[7] 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.[7] Twenty-five percent of patients had masses discovered incidentally during a routine examination or abdominal surgery (cholecystectomy). The study done by Cody *et al*. reported abdominal pain and weight loss to be the most common symptoms.[8] The above symptoms are at par with our case. 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.[7] Leiomyosarcoma on CT is typically nonfatty, irregularly marginated, heterogeneous mass. Ultrasound may be

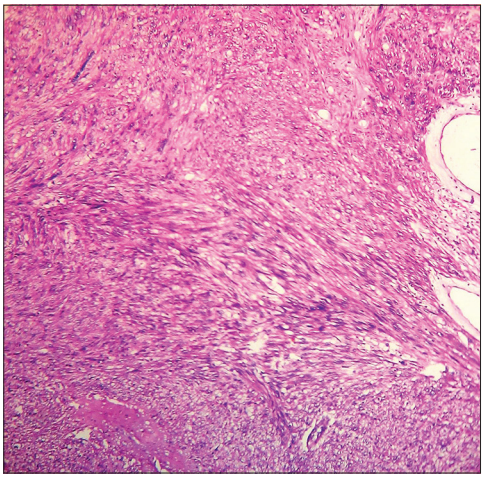

Figure 2: Micrograph showing spindle cells arranged in nodules and fascicles (H and E, ×20)

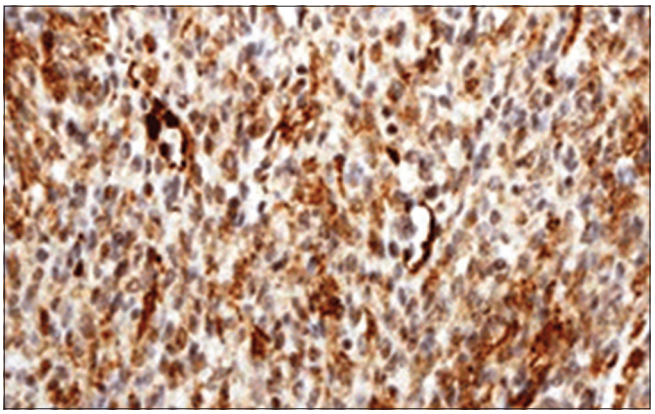

Figure 4: Micrograph showing diffuse smooth muscle actin positivity

useful in delineating tumor vessels or vascular infiltration. Surgery is the main treatment factor in the outcome of the retroperitoneal leiomyosarcoma.[9,10] Complete surgical resection with at least 3 cm margins is the treatment of choice but is rarely feasible due to invasion of adjacent structures by the tumor. Curative surgery is difficult with large retroperitoneal sarcomas and those in close proximity to vital structures and involving adjacent organs.[9]

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Cite this article as: Sawaimul K, Iqbal BM, Kambale T. Retroperitoneal leiomyosarcoma in a young adult presenting as a huge abdominal mass. Clin Cancer Investig J 2015;4:558-60.

Source of Support: Nil, **Conflict of Interest:** None declared.