Primary Leiomyosarcoma of Spleen Masquerading Adrenal Tumor - A Rare Case

Abstract

Leiomyosarcoma (LMS) is a malignant smooth muscle tumor which is predominantly intra-abdominal in location. LMS arising from vascular smooth muscle is quite uncommon which usually arises from inferior vena cava and is aggressive in nature. Primary venous LMS of splenic vein is exceedingly rare and has unpredictable clinical course with complete surgical resection representing the only potentially curative treatment. We hereby report a case of primary splenic vein LMS in a 59-year-old male patient mimicking an adrenal tumor radiologically.

Keywords: *Leiomyosarcoma, rare, spleen*

Zeenat Ara, Piyabi Sarkar[1], Keya Basu, Probhas Kumar Sarkar[2]

Departments of Pathology and [2]General Surgery, Institute of Post Graduate Medical Education and Research, [1]Department of Pathology, Nil Ratan Sircar Medical College and Hospital, Kolkata, West Bengal, India

Introduction

Leiomyosarcoma (LMS), a malignant mesenchymal tumor, is derived from smooth muscle and commonly found in the intra-abdominal locations such as retroperitoneum, mesentery, or omentum as well as in deep soft tissues of extremities.[1] Approximately 2% of LMSs arise from vessel wall smooth muscle and veins are more frequently affected than arterioles.[2] Tumors arising from inferior vena cava constitute majority of the cases and are predominantly found in middle-aged female patients.[1] LMS derived from splenic vein is extremely rare with only four cases reported in the literature.[2-5] Therefore, we report a case of splenic vein LMS in a 59-year-old male patient simulating an adrenal neoplasm.

Case Report

A 59-year-old male patient presented with the chief complaints of low-grade fever, left-sided abdominal pain and fullness of flanks for the past 2 months. He was a known hypertensive and on antihypertensives since the past 5 years. There was no history of any other comorbidity.

On examination, he was alert, conscious, and pale (pallor 3+) with a heart rate of 104/min and a blood pressure of 130/80 mm Hg. His abdomen was distended more on the left side, and a palpable mass was detected spanning across left hypochondrium, left lumbar and umbilical region, and extending 7 cm below the left costal margin.

Routine investigations revealed, hemoglobin 5.6 g/dl, total leukocyte count 9600/cumm, and platelet count 2.65 lacs/cumm. Serum electrolytes, renal and liver function tests were normal. Chest X-Ray showed left-sided pleural effusion. Ultrasonography of the whole abdomen revealed a huge lesion, measuring 12.26 cm × 10.12 cm, in left hypochondrium and left lumbar region with heterogeneous echotexture. Colour Doppler suggested an increased blood flow pattern. A contrast-enhanced computed tomography (CECT) scan was done which suggested a heterogeneously enhancing soft tissue mass with focal areas of calcification located in the left suprarenal region, measuring about 16.9 cm × 15.5 cm × 10.9 cm, and pushing the kidney medially and downward [Figure 1]. The overall impression on CECT was that of a large left adrenal neoplastic mass with left-sided moderate pleural effusion.

With the presumption of an adrenal mass, further tests were performed such as plasma metanephrine, urinary vanillylmandelic acid, urinary metanephrine, serum LDH, and serum uric acid, but all were found to be within normal limits. The patient was planned for a laparotomy.

On laparotomy, a large tumor mass was noted and on careful dissection, it was

How to cite this article: Ara Z, Sarkar P, Basu K, Sarkar PK. Primary leiomyosarcoma of spleen masquerading adrenal tumor - A rare case. Clin Cancer Investig J 2017;6:200-2.

Address for correspondence:
Dr. Zeenat Ara,
First Floor, Junaid Enclave, Jagannathpur, Near Shimultala Football Ground, Kolkata - 700 150, West Bengal, India.
E-mail: ara.zeenat7@gmail.com

Access this article online

Website: www.ccij-online.org

DOI: 10.4103/ccij.ccij_40_17

Quick Response Code:

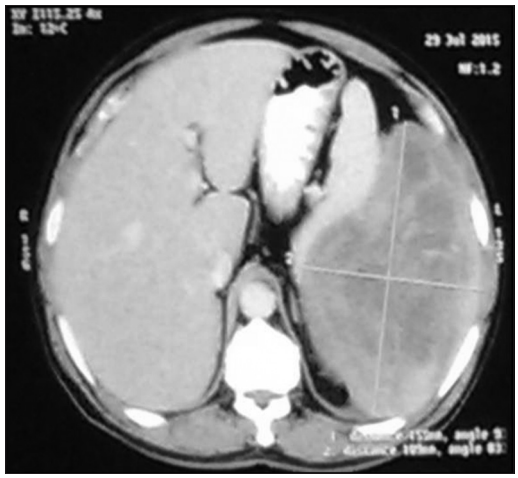

Figure 1: Contrast-enhanced computed tomography of abdomen showing a heterogeneously enhancing soft tissue mass (m-16.9 cm × 15.5 cm × 10.9 cm) with focal areas of calcification located in the left suprarenal region and pushing the kidney medially and downward

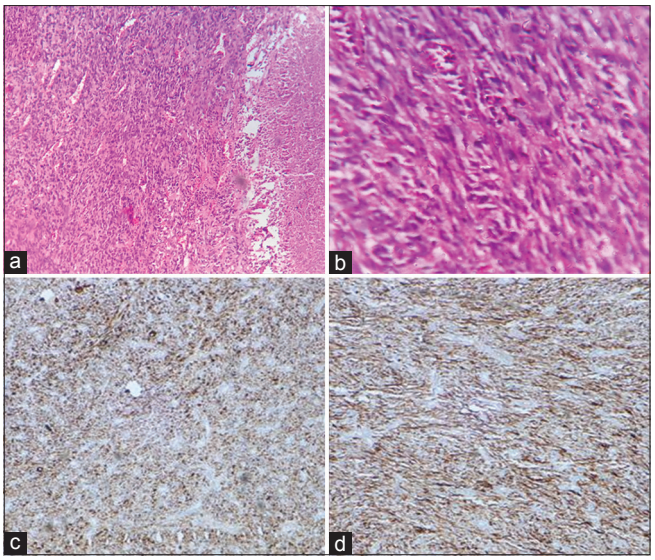

Figure 2: (a) Section from tumor mass showing atypical spindle-shaped cell population with areas of extensive necrosis (H and E, ×100). (b) Higher magnification showing pleomorphic, plump, spindle-shaped tumor cells arranged in intersecting fascicles with focal hemangiopericytoma-like arrangement (H and E, ×400). (c) Diffuse positive staining for smooth muscle actin (×400). (d) Diffuse positive staining for Desmin (×400)

found to be arising from the lower pole of spleen and adjoining areas of splenic hilum. Thus, a splenectomy with partial omentectomy was done and the specimen was sent to our Department of Pathology for further investigation.

On gross examination, a huge tumor mass measuring, 35 cm × 24 cm × 12 cm, attached to the hilum of spleen was noted. Cut section was variegated in appearance with hemorrhagic and necrotic areas. Part of omentum sent separately measured 40 cm × 16 cm × 1 cm. No lymph nodes were identified.

On histopathological examination, several sections from different parts of tumor showed the histological features of a spindle cell tumor composed of tumor cells with moderate amount of cytoplasm with indistinct cell borders, plump spindle-shaped pleomorphic nuclei. Numerous atypical mitotic figures (8–9/10 high power field) were present along with extensive areas of necrosis (<50% of tumor mass) [Figure 2a and b] and mixed inflammatory cell infiltrate. Several sections from omentum showed fibrofatty tissue within normal limits.

On immunohistochemistry, the tumor cells were positive for smooth muscle actin and Desmin [Figure 2c and d], while they were negative for CD117 and DOG1, thus ruling out a diagnosis of extraintestinal gastrointestinal stromal tumor.

The tumor was given the diagnosis of primary LMS of spleen, Grade 1 tumor, according to the FNCLCC classification.[6,7]

Postoperatively, the patient was severely hypotensive but was resuscitated with crystalloids and vasopressors. He improved gradually and was discharged on the 10th postoperative day. Follow-up fluorodeoxyglucose positron emission tomography (PET) scan [Figure 3] done 3 weeks later showed the presence of mildly hypermetabolic heterogeneously enhancing soft tissue mass lesion with cystic areas in the splenic fossa extending up to the upper two-third of left kidney causing indentation of left renal cortex which was suggestive of residual sarcomatous disease.

Discussion

LMS, a rare malignant tumor of smooth muscle origin is usually located in the uterus, retroperitoneum, subcutaneous tissue or deep soft tissue with only 2% of all cases originating from blood vessels.[1] 75% of large-vessel LMSs arise from inferior vena cava, although other retroperitoneal veins such as renal, iliac, ovarian, and spermatic veins may also be affected.[3]

Splenic vein LMS is an extremely rare finding, and to the best of our knowledge, only four such cases have been reported so far. The first case of splenic vein LMS involving the pancreatic tail and the splenic hilum was reported by Rödl and Hofmann-Preiss[5] Three more cases were reported by Gage *et al*.[1] (also reported by Niver *et al*.[4]), Aguilar *et al*.[3] and Patrono *et al*.[2] respectively.

Primary venous LMS is a potentially aggressive disease with a 5-year survival rate of 33%–68%. It is commonly found in women in the sixth decade of their life. The tumor remains asymptomatic for a prolonged period of time and usually attains considerable size at the time of diagnosis.[2] The most important criteria for malignancy are large size, high cellularity, nuclear pleomorphism, increased number of atypical mitotic figures and distant spread.[8]

The important differential diagnosis includes metastasis, extraintestinal gastrointestinal stromal tumor, Schwannoma, inflammatory myofibroblastic tumor, and fibrous solitary tumor.[2]

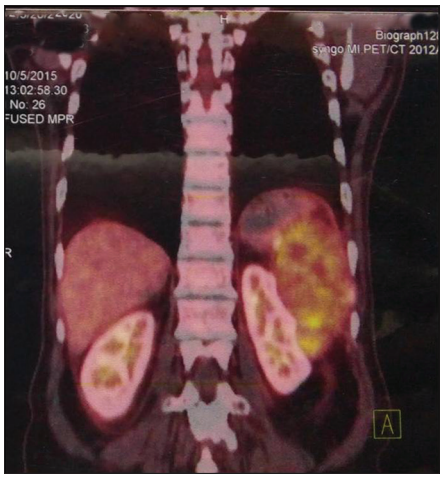

Figure 3: Fluorodeoxyglucose positron emission tomography scan showing a mildly hypermetabolic, heterogeneously enhancing soft tissue mass lesion with cystic areas in the splenic fossa, extending up to the upper two-third of left kidney causing indentation of left renal cortex, suspected residual tumor

The ideal treatment protocol for splenic LMS is complete *en bloc* surgical resection of all involved organs.[9,10] In the absence of overtly disseminated disease, resection usually provides the opportunity for complete cure.[1] The value of postoperative radiation therapy as well as chemotherapy is controversial but due to high risk of local recurrence, adjuvant radiation therapy is often employed. Patients who are unfit for resection usually receive palliative therapy, and they generally have poor prognosis.[2] In our case, residual sarcomatous tumor was detected on PET-scan, and the patient was planned for adjuvant radiation therapy on follow-up.

Conclusion

Owing to rarity of primary splenic vein LMS, a definite treatment protocol is yet to be established. Radical surgical resection with close monitoring of patients is the treatment mainstay.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Gage MJ, Newman E, Maldonado TS, Hajdu CH. Leiomyosarcoma of the splenic vein. J Vasc Surg 2012;55:1485-7.
2. Patrono D, Molinaro L, Mazza E, Romagnoli R, Salizzoni M. Splenic vein leiomyosarcoma: Case report and review of the literature. JOP 2014;15:512-4.
3. Aguilar C, Socola F, Donet JA, Gallastegui N, Hernandez GA. Leiomyosarcoma of the splenic vein. Clin Med Insights Oncol 2013;7:263-8.
4. Niver BE, Megibow AJ, Faust MJ, Rosenkrantz AB. Multidetector CT appearance of leiomyosarcoma of the splenic vein. Clin Radiol 2011;66:688-90.
5. Rödl W, Hofmann-Preiss K. Leiomyosarcoma of the splenic vein. Radiologe 1988;28:42-4.
6. Guillou L, Coindre JM, Bonichon F, Nguyen BB, Terrier P, Collin F, *et al.* Comparative study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group grading systems in a population of 410 adult patients with soft tissue sarcoma. J Clin Oncol 1997;15:350-62.
7. Miettinen M. Immunohistochemistry of soft tissue tumours - Review with emphasis on 10 markers. Histopathology 2014;64:101-18.
8. Golden T, Stout AP. Smooth muscle tumours of the gastrointestinal tract and retroperitoneal tissues. Surg Gynecol Obstet 1941;73:784-810.
9. Hollenbeck ST, Grobmyer SR, Kent KC, Brennan MF. Surgical treatment and outcomes of patients with primary inferior vena cava leiomyosarcoma. J Am Coll Surg 2003;197:575-9.
10. Mingoli A, Cavallaro A, Sapienza P, Di Marzo L, Feldhaus RJ, Cavallari N, *et al.* International registry of inferior vena cava leiomyosarcoma: Analysis of a world series on 218 patients. Anticancer Res 1996;16:3201-5.