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Case Report

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LEIOMYOSARCOMA OF THE SPLENIC VEIN: RARE ENTITY IN UNCOMMON LOCATION

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ABSTRACT

Leiomyosarcoma (LMS) is a malignant smooth muscle tumour with those arising from the vascular smooth muscle being quite uncommon. Primary leiomyosarcoma of splenic vein is exceedingly rare and has unpredictable clinical course with complete surgical resection representing the only potentially curative treatment. We report a case of primary splenic vein LMS adjoining the splenic hilum in a 55-year old male who presented with complaints of left sided abdominal pain.

KEYWORDS: MDCT, Leiomyosarcoma, splenic vein.

INTRODUCTION

Leiomyosarcoma(LMS) is a malignant smooth muscle tumour, most commonly found in the abdomen or in the limbs. Those involving the vessels, commonly the veins arise directly from their muscular walls.[1] LMS arising from the splenic vein is exceedingly rare with only few case reports published in the literature.[3-7] We hereby present a case of a primary splenic vein LMS arising in the perihilar region in a 55 year old male patient.

CASE REPORT

A 55 year old hypertensive male presented with complaints of recurrent left flank pain and nausea, unrelated to meals. Laboratory investigations including tumour markers and upper gastrointestinal endoscopy did not reveal any significant abnormality. An ultrasound was suggestive of a splenenculus at the splenic hilum with no other abnormality.

Hence, a contrast enhanced computed tomography (CT) scan of the abdomen and pelvis was performed for further evaluation on a 64 slice Siemens Somatom cardiac machine with bolus tracking technique acquiring arterial and venous phase images. A relatively well demarcated lobulated nodular soft tissue mass lesion with heterogenous post contrast enhancement was seen in the perisplenic region. It was superior to the splenic hilum, focally abutting and indenting the splenic parenchyma, greater curvature of stomach and pancreatic tail, encasing few twigs of the splenic artery and compressing and appearing inseparable from the distal splenic vein. No abnormality was detected in rest of the abdomen and pelvis. The possible differentials included a splenic hamartoma, lymphoma and gastrointestinal stromal tumour.

A CT guided biopsy was performed, which on histopathology was suggestive of a smooth muscle tumour. The patient then underwent an exploratory laparotomy with distal pancreatectomy and splenectomy along with resection of the lesion. Intra-operatively the mass was seen adherent to the spleen, centered at the splenic hilum and was reaching upto the distal pancreas. The patient had an uneventful post operative recovery.

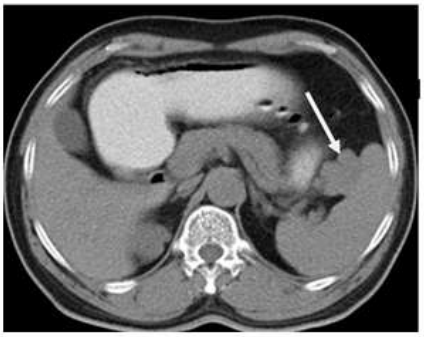

Figure 1(a)

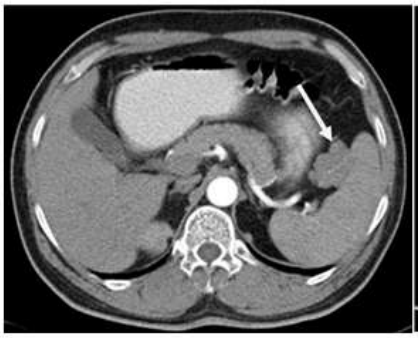

Figure 1(b)

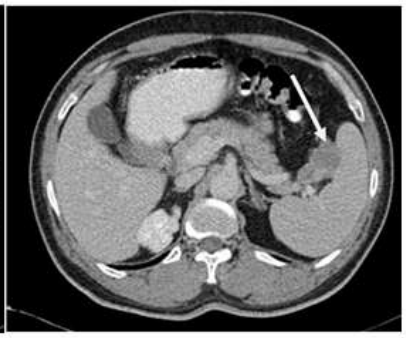

Figure 1(c)

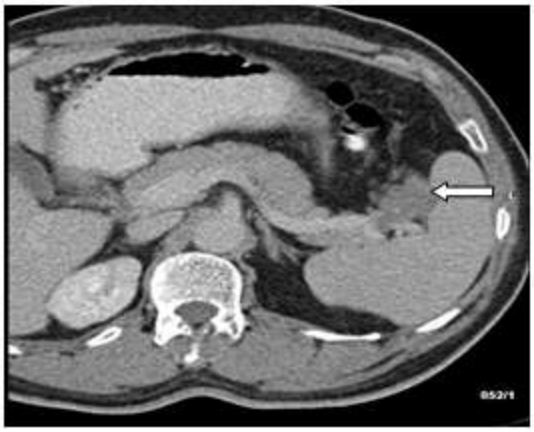

Figure 1(d).

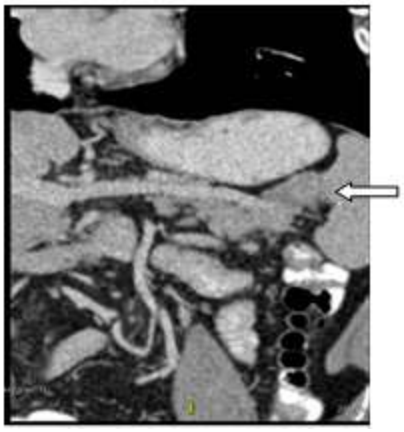

Figure 1(e).

Figure 1(a): Non contrast enhanced CT, 1(b): contrast enhanced CT: arterial phase, 1(c): Contrast enhanced CT venous phase: showing a heterogenously enhancing lobulated nodular soft tissue lesion in the perisplenic region. Figure 1(d) and 1(e) curved MPR images of the same patient.

The cut specimen revealed a firm white nodular mass with a whorled appearance abutting the splenic hilum and pancreatic tail with no obvious infiltration of the same.

Figure 2: Gross specimen: Cut surface of tumour showing white and whorled appearance.

Final histopathology was suggestive of a LMS arising from the splenic vein. Immunohistochemistry staining was positive for smooth muscle actin and Desmin, and for S100, CD117 & CD31. Pan-cytokeratin was negative. Mitotic index was 8/10 HPF.

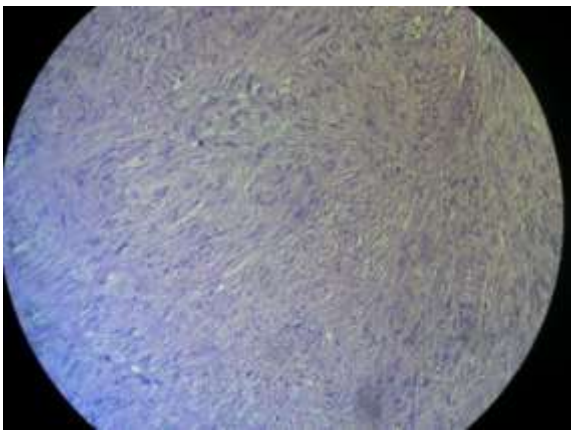

Figure 3: Hematoxylin eosin staining showing spindle cells in dense fibrous stroma.

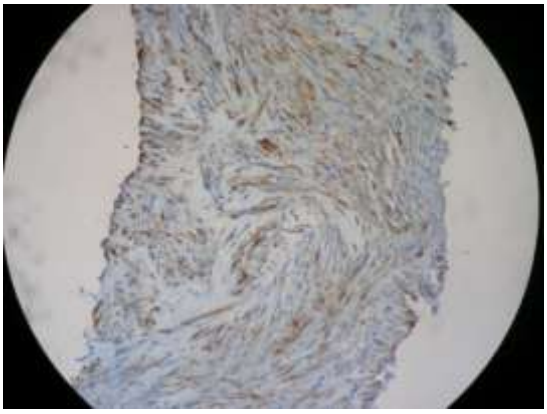

Figure 4(a).

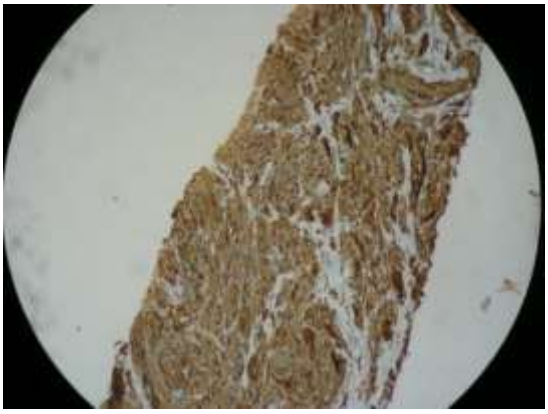

Figure 4(b).

Figure 4 (a) and (b): Immunohistochemistry staining showing desmin and smooth muscle actin positivity.

DISCUSSION

LMS, a rare malignant tumour of smooth muscle origin, is usually located in the uterus, retroperitoneum, subcutaneous tissues or deep soft tissues with only 2% of all cases originating from blood vessels.[1,2] 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.[11]

Similar to other primary venous LMSs, those involving the splenic vein seem to occur mainly in women in their sixth decade of life.[17] In our case, the patient was a 55 year old male.

Splenic vein LMS is an extremely rare lesion, and to the best of our knowledge, only few such cases have been reported so far in the literature. The first case of splenic vein LMS involving the pancreatic tail and the splenic hilum was reported by Rödl and Hofmann-Preiss,[3] four more cases were reported by Gage et al. (also reported by Niver et al.), (mixed attenuation mass along the posterior aspect of the pancreatic body), Aguilar et al. (solid heterogeneous 12 × 10 cm mass located in the tail of the pancreas displacing the stomach and compressing the left kidney), Patrono et al (15 mm mainly hypodense lesion along the posterior margin of the pancreatic body), &Wenfangwu et al (irregular, hypodense mass measuring ~4.1x3.0 cm in the tail of the pancreas) respectively.[4-7,11] In all the reported cases the tumour was located in the distal pancreas and pancreatic tail.

Regarding imaging techniques, ultrasound is often the first diagnostic tool because it does not expose patients to ionizing radiation, and it is widely available. However, the accuracy of ultrasound for diagnosing tumours is low.[12] For that reason, CT scan and magnetic resonance imaging (MRI) are very important because they allow us to determine the extension of the tumour to surrounding organs and the presence of vein thrombosis and distant metastases.[13] A highly sensitive and specific modality for detecting recurrence in post-therapy patients with sarcoma is 18F-fluorodeoxyglucose (FDG) positron emission tomography (PET-CT). However, it provides no significant advantage over CT scan or MRI for this purpose.[14]

Surgical resection remains the mainstay of treatment for vascular LMS. The ideal treatment protocol for splenic vein LMS is complete en bloc resection of all involved organs with adjuvant treatment depending upon the metastatic potential of sarcoma.[2,8,10,16,18] Resection offers the only opportunity for complete cure in the absence of disseminated disease. In addition to tumour extent, histologic evaluation for classification and grade is also important in guiding treatment. While other retroperitoneal sarcomas are plagued by local recurrences, vascular LMS most often recur at distant sites, with reported metastatic rates of ~50% with the lung and liver being the most common sites.[9,11,15] A careful post-surgical follow-up is warranted for these patients.

Our patient underwent radical surgery and no adjuvant chemotherapy or radiation therapy was therefore given. He is currently on follow up at our institution.

CONCLUSION

LMS of the splenic vein is an extremely rare entity. Due to very few cases being reported, enough studies and trials are not available regarding clinical course, imaging findings, treatment protocols and the prognosis. A tumour in the splenic hilum or pancreatic tail with nonspecific findings should include a differential diagnosis of LMS. Because of the insidious evolution of this neoplasm, follow-up imaging studies should be exhaustive, and are even more necessary than in other tumour types, to find out local recurrence or distant metastases

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