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CASE REPORT

Leiomyosarcoma arising from the inferior mesenteric vein draining in the splenomesenteric angle with a tumour thrombus at the splenomesenteric confluence: a case report and review of the literature

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Introduction

Leiomyosarcoma (LMS) is a very rare vascular tumour that arises from the media of the blood vessels. We present the case of a 64-year-old patients affected by LMS of the inferior mesenteric vein (IMV) with a tumour thrombus treated with surgery and adjuvant chemotherapy. In this case report, we want to point out the importance of a dedicated multidisciplinary soft tissue sarcoma team in planning the right surgical strategy.

Case presentation

A 64-year-old man presented to his primary care physician with a 6-week history of general discomfort, abdominal bloating and increased intestinal transit. Physical examination was negative except for light epigastric pain to deep abdominal palpation. Peripheral blood count and biochemical test results were normal. Abdominal ultrasonography was negative. Esophagogastroduodenoscopy and colonoscopy were negative. Abdominal computer tomography showed a 10-cm lobulated mass in the left flank in contact with the Treitz loop. The mass infiltrated the IMV that showed an intraluminal thrombus protruding into the splenomesenteric angle (Fig. 1). The venous phase showed huge gastroepiploic shunts. Histopathological examination after a CT-guided biopsy revealed a spindle cell sarcoma with myogenic differentiation (reactivity for smooth actin and h-caldesmon on immunohistochemistry). Laparotomy confirmed the presence of a lobulated solid mass infiltrating the first jejunal loop. Manual palpation of the liver detected a solid 1-cm sub-glissonian lesion in segment VI. A limited resection was performed to remove the hepatic lesion. No other hepatic lesions were detected. To detach the mass from the first jejunal loop, a tangential resection of the jejunum was performed. Once the mass was mobilised from the surrounding tissues, it was clear that the tumour originated from an anatomical variant of the IMV draining in the splenomesenteric angle. The superior mesenteric vein and the splenic vein were isolated and temporarily clamped, the IMV was sectioned proximally to the thrombus and the vein was sutured with a transversal continuous polypropylene suture. After the IMV was sectioned, we observed a transient congestion of the sigmoid colon that resolved spontaneously, probably due to the gastroepiploic shunts previously seen on the CT scan. The postoperative course was uneventful. The patient was discharged on the sixth postoperative day. Pathological examination of the surgical specimen confirmed the diagnosis of low-grade LMS of vascular origin (D1 N1 M1, grade I according to the Fédération Nationale des Centres de Lutte Contre le Cancer classification), characterised by foci of coagulative necrosis (20 %), immunoreactivity for myogenic markers (smooth muscle actin, h-caldesmon and negativity for Desmin, C-kit and CD 34−) and a low number of mitosis (3 × 10HPF) (Fig. 2a–c). Furthermore, a spindle cell sarcoma was also documented in the hepatic nodule, indicating a metastasis from the primary LMS

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(Fig. 2d). The jejunal loop was not infiltrated. No adjuvant therapy was performed. A radiological follow-up was planned. Three months later, the patient underwent a CT scan that was negative for local and distant metastases. Six months later, an abdominal CT scan detected one focal liver lesion in segment 6. Four more liver lesions were detected during ultrasound-guided fine needle biopsy and histopathological examination revealed the presence of LMS metastases. The patient was treated with anthracycline-based adjuvant chemotherapy. The patient is alive 13 months after the original procedure with stable disease.

Discussion

We report the third case of LMS of the IMV treated with surgery. Radical surgical excision is the gold standard treatment for retroperitoneal and visceral abdominal soft tissue sarcomas [1–3], but a multidisciplinary approach is crucial to plan the right therapeutic strategy.

LMS of vascular origin are rare tumours that arise from the media of blood vessels, with venous tumours being five times more common than tumours of arterial origin [4]. The inferior vena cava is the most common site of origin reported for venous LMS, followed by major veins of the extremities, iliac, saphenous, renal, pulmonary, axillary, femoral, internal jugular, uterine, antecubital, azygous, hepatic and ovarian veins. In 1947, an LMS arising from the inferior left colic vein was resected by Puig Sureda in a 61-year-old woman [5]. The first report of a venous LMS arising from the IMV is the case reported by Chorquat in 1976 [6]. He reported the case of an 83-year-old patient with solid mass arising from the external wall of the IMV. The IMV was sectioned and ligated. No vascular congestion of the left colon was observed. Histopathology revealed a vascular LMS. The second case of LMS arising from the IMV is the one reported by Clemente et al. in 2009 [7]. They reported the case of a 66-year-old woman with an LMS of the IMV that underwent segmental resection of the vein, including the mass, and left colonic resection. The patient is disease free 24 months after surgery. Only four cases of LMS of the portal system have been reported in scientific literature, two of which originated from the superior mesenteric vein [8–10]. In these cases, the mesenteric flow was restored with an autologous saphenous graft. In our case, we did not need to restore the mesenteric flow after the IMV transection due to the presence of huge venous gastroepiploic shunts.

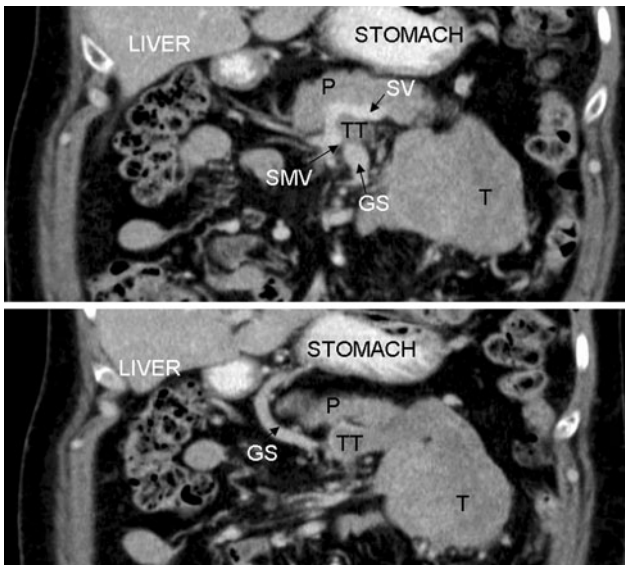

Fig. 1 Preoperative CT scan showing tumour thrombus (*TT*) protruding into the splenomesenteric angle. Superior mesenteric vein (*SMV*), splenic vein (*SV*), gastroepiploic shunt (*GS*), pancreas (*P*) and main tumour lesion (*T*)

In this case report, we show how the presence of a multidisciplinary soft tissue sarcoma team is extremely important to plan the right therapy. Preoperative pathological examination was important for two reasons. The first is that no neoadjuvant treatment was performed because preoperative biopsy showed a low-grade LMS; besides, there was no evidence in scientific literature that neoadjuvant chemo and radiation therapy could affect overall survival. The second reason was that once the vascular origin of the lesion was determined, we were able to perform tangential resection of the duodenal loop with safe surgical margin. An experienced radiological team and a new-generation CT scan were crucial to evaluate vascular anatomy and to perform the extension of the tumour thrombus and percutaneous biopsy. IMV draining in the splenomesenteric angle is an anatomical variant reported in 18 % of patients [11]. Preoperative detection of gastroepiploic shunts has been important to foresee the left colon transient congestion.

Various chemotherapy regimens, such as doxorubicin, trabectedin, gemcitabin, ifosfamide, cyclophosphamide and vincristine, have been proposed for venous LMS, although no extensive clinical studies using these regimens have been documented. Currently perioperative chemotherapy with adjuvant intent is not the standard treatment for primary tumours. Also in the metastatic setting, its use as complementary treatment is controversial in case of resectable limited metastatic burden. Radiation therapy is often used to improve local control, but there is no evidence in literature that this can have any clinical benefit [12]. In our case, we did not offer postoperative treatments because the patients had a single, small metastasis

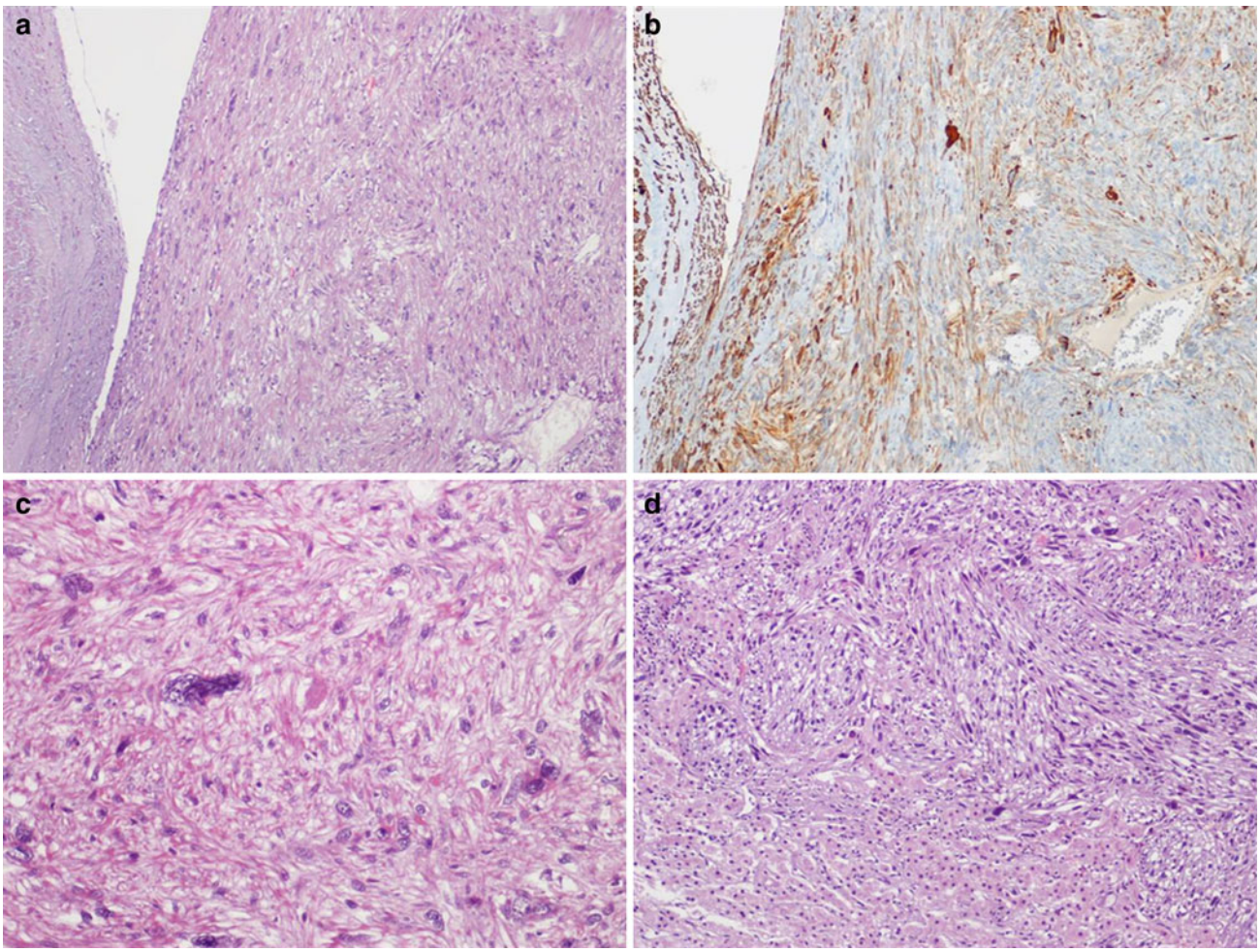

Fig. 2 **a** Leiomyosarcoma of the mesenteric vein. Tumour protrudes and partially occludes the lumen (E/H ×100). **b** h-Caldesmon immunoreactivity underlines the myogenic origin of this spindle cell sarcoma. Note the residual vascular wall on the *left* (IHC ×100). **c** Higher magnification showing scattered atypical cells in a background of more classic fascicular leiomyosarcoma (E/H ×200). **d** Liver metastasis from vascular leiomyosarcoma which recapitulates the primary tumour (E/H ×100)

from a low-grade tumour. The patient was treated with chemotherapy after the evidence of nonresectable metastatic recurrence, according to ESMO guidelines.

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Conflict of interest The authors declare that they have no conflict of interest.

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