

Leiomyosarcoma of the superior mesenteric vein: a diagnostic and therapeutic challenge

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Abstract

We report here a case of a very rare entity, a leiomyosarcoma of the mesentericoportal trunk, which was initially misdiagnosed as an unresectable pancreatic cancer invading the mesenteric vein, and which was finally treated by pancreatectomy with mesentericoportal reconstruction. The pitfalls of diagnosis and modalities of resection are discussed.

Key words Leiomyosarcoma · Superior mesenteric vein · Whipple procedure · Gore-Tex prosthesis

Case report

Because of sudden epigastric pain, a 50-year-old woman underwent an abdominal ultrasound, revealing a 3 cm-large tumor in the inferior part of the head of the pancreas, with possible invasion of the mesenteric vein. She had no past medical or surgical history and her general status was remarkably good. Laboratory tests and, especially her liver profile, were normal. An abdominal computed tomography scan confirmed the existence of a hypodense mass with severe compression of the distal section of the mesenteric vein, which was not occluded (Fig. 1). No dilatation of the bile duct, or the pancreatic duct, nor metastasis was observed.

Endoscopic ultrasound was performed, in order to precisely determine the vascular involvement and to perform a secure biopsy.^{1,2} This imaging confirmed a solid and cystic mass with mesenteric vein invasion. Biopsies found spindle cells, suggesting sarcoma, the origin of which could not be determined.

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Resection was then evaluated: a visceral arteriogram revealed a normal superior mesenteric artery, but showed that the mesenteric vein was not occluded, although it had been invaded to a relatively short length (Figs. 2 and 3).

Pancreato-duodenectomy (Whipple procedure) with en-bloc resection of the spleno mesenterico-portal venous confluence was done in order to perform a large and complete R0 resection. The defect after resection was 7 cm in length, so that it was impossible to perform end-to-end mesentericoportal reconstruction even after mobilization on the mesentery. So, portal revascularization was done, with a Gore-Tex (BARD® IMPRA® tempe, AZ, USA) prosthesis (length, 7 cm; diameter, 8 mm; Fig. 4). Apart from the drainage of an infected collection, the postoperative course was uneventful.

Final pathology confirmed the diagnosis of leiomyosarcoma of the mesenteric vein. Indeed, spindle cells were found within the smooth muscle fibers infiltrating the wall of the mesenteric vein and the nearby pancreas. Surgical margins were free from tumor. At the time of writing, 18 months after surgery, the patient is free of recurrence. She is still on antiaggregant treatment and has a permeable Gore-Tex prosthesis.

Discussion

Leiomyosarcomas of vascular origin are rare tumors, which arise from the media of blood vessels. They occur predominantly in women. Tumors of venous origin are five times more common than arterial ones, with the inferior vena cava being the most common origin. Concerning leiomyosarcoma of the portal venous system, the literature is very sparse: only five previous cases have been reported in the English language literature.³⁻⁷ Our patient is the first to be treated by a Whipple procedure combined with a vascular reconstruction using a Gore-Tex prosthesis.



Fig. 1. Computed tomography scan of abdomen, demonstrating a 30 × 37 × 20-mm mass with compression/or invasion of mesenteric vein



Fig. 2. Preoperative arteriography

Two recent published cases report leiomyosarcoma of the portal venous system with two different treatments. The first patient was a 66-year-old man suffering from a leiomyosarcoma of the mesenteric vein, requiring a right hemicolectomy with an autogenous saphenous vein graft.⁶ The other patient, a 40-year-old woman, was operated on for a leiomyosarcoma arising from the superior mesenteric vein, which was revealed by a mass originating from the head of the pancreas.⁷ In this patient, the tumor could be separated from the head of the pancreas, and vein resection with saphenous graft reconstruction was performed. The follow-up for both



Fig. 3. Preoperative indirect mesenteric portogram

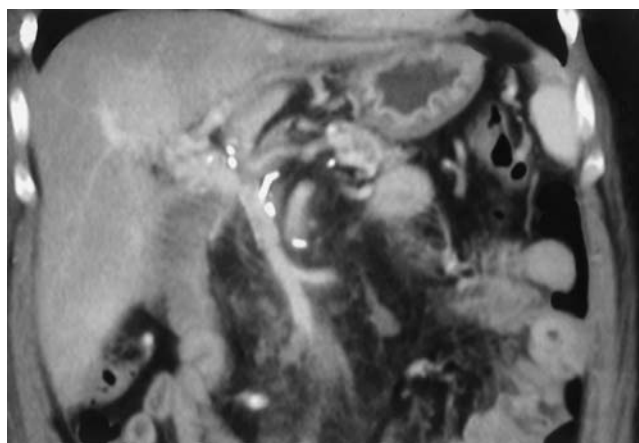


Fig. 4. Postoperative computed tomography scan

patients was short (6 and 12 months), with, for one patient, a liver recurrence 6 months after surgery.

Our experience confirms that a complete surgical resection is possible, requiring vascular reconstruction, and in that location, a Whipple procedure, with good results. Such extended surgery is, today, routinely and safely performed in pancreatic cancer, provided that R0 resection can be achieved.⁸

The challenge for these lesions is, first, diagnostic: indeed, a pancreatic tumor was our first diagnosis. But this tumor was more regular, and limited, and not infiltrating, such as can be seen in front of a pancreatic cancer this size.

From a therapeutic point of view, in this case, as in all sarcomas, surgery is the only chance of cure, and large resection is often required. Whipple resection, in our patient, was performed in order to obtain tumor-free resection margins. Pathology retrospectively justified the pancreatic resection, showing the infiltration of the pancreatic tissue. The modality of vascular reconstruction can also be discussed: a homograft is a good option, lowering the risk of secondary thrombosis. But, because of the large tumor bed due to the Whipple procedure and because of the long defect (more than 6 cm),⁸ we thought that a Gore-Tex prosthesis would be better and thus avoid a venous plication. In our experience this vascular reconstruction is still permeable 1 year later.

Leiomyosarcoma of the mesentericoportal venous system is a very rare entity mimicking a pancreatic mass. An aggressive surgical attitude must be adopted, with possible Whipple procedure associated with vascular reconstruction. A Gore-Tex prosthesis can be used with good results, especially when the defect is long.

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