SHORT REPORT

A Rare Tumour at an Unusual Site; 6 Year Survival Following Multiple Liver Resections for Metastatic Leiomyosarcoma of the Superior Mesenteric Vein

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Leiomyosarcoma of the superior mesenteric vein has been reported only once before. Aggressive surgery is the primary therapy and main prognostic factor in leiomyosarcoma. The 5 year survival remains poor at around 50% for a wide range of sarcomas. We present a case of metastatic leiomyosarcoma of the superior mesenteric vein that resulted in multiple hepatic resections and six year survival.

Case Report

A 43-year-old lady presented with a 3 month history of periumbilical pain. She had previously had a normal diagnostic laparoscopy for lower abdominal pain some years ago.

On examination she had a slightly distended epigastrium and a sausage-shaped mass in the left upper quadrant. An OGD and a colonoscopy were both normal and an abdominal ultrasound demonstrated a slightly lobulated, mobile, solid mass lying in the left upper quadrant, that did not appear to be related to any particular abdominal organ. She underwent an abdominal CT scan with oral contrast. This showed a mass which displaced the upper small bowel loops but there was no relation to the transverse colon. The liver, spleen, kidneys, and pancreas were all normal. A small bowel meal was normal.

At laparotomy, a nodular solid mass was found arising from the root of the small bowel mesentery at the level of the duodeno-jejunal flexure. There was no evidence of disease elsewhere and with further dissection the mass was found to arise from the main trunk of the superior mesenteric vein with no other connections and no plane of cleavage between the tumour and the vein. Following the administration of 5000 units of heparin intravenously, the specimen was excised en-bloc with 2 cm of superior mesenteric vein and a primary end-to-end anastomosis was carried out with good flow in the vein afterwards. An anastomosis was performed on safety grounds as ligation of the Superior Mesenteric Vein may have led to small bowel venous infarction. The patient made an uneventful recovery and was discharged after 6 days.

The histology demonstrated irregular broad fascicles of smooth muscle cells with moderate nuclear pleomorphism and variable cellularity. The tumour cells showed strong positivity with actin and desmin confirming smooth muscle origin. The features were consistent with a leiomyosarcoma arising from, and partially replacing the wall of the superior mesenteric vein. The vein margins were clear of tumour by a narrow margin and the main tumour mass was covered by a thin pseudo capsule of connective tissue.

The patient went on to have 6 cycles of Adriamycin chemotherapy but one year later surveillance CT revealed a solitary liver metastasis for which central hepatectomy was performed. There was no evidence of local recurrence. The following year 3 more
deposits were removed at segmental hepatectomy, again histology confirming metastatic leiomyosarcoma. Three years after her initial operation she underwent a completion right hemihepatectomy for further metastatic disease leaving just segments I, II and III.

Five years after initial diagnosis CT revealed multiple lung lesions which were considered irresectable. Now in her sixth year after diagnosis she has chest and abdominal wall secondaries and is being treated palliatively.

Discussion

Leiomyosarcoma may arise from virtually any site but there has only been one previous report of one arising from the superior mesenteric vein.\(^1\) Classification of these tumours is difficult and it has been proposed that abdominal and retroperitoneal leiomyosarcomas which are not directly associated with the gastrointestinal tract be designated as extra-gastrointestinal stromal tumours. In this case the tumour was clearly arising from the wall of the superior mesenteric vein rather than from an unspecified area of the retroperitoneum and should probably be classified as venous in origin rather than retroperitoneal.

Series relating to leiomyosarcoma of the inferior vena cava\(^2\) and to gastrointestinal stromal tumours\(^3\) have both emphasized the importance of complete resection of the primary tumour in terms of outcome although grossly clear margins rather than microscopically clear margins appears to be important.\(^3\) This can obviously pose difficulties when essential structures such as the superior mesenteric vein are involved. Techniques developed from pancreatic surgery and from transplantation allow resection of a segment of superior mesenteric vein with either primary anastomosis or a vein graft with internal jugular vein to ensure any anastomosis is tension free (Fig. 1).

Aggressive surgery is the primary therapy and main prognostic factor in leiomyosarcoma although outcome remains poor with 5 year survival of around 50% for a wide range of sarcomas.\(^2\) This has stimulated interest in the use of adjuvant therapy but as yet none has demonstrated a definitive benefit which is likely to remain a problem in view of the scarcity of the tumour. Systemic chemotherapy for retroperitoneal sarcomas has so far failed to demonstrate any benefit although there is evidence that radiotherapy reduces local recurrence. Liver resection for the treatment of metastatic leiomyosarcoma is proven to prolong survival\(^4\) and 5-year survival has been reported.\(^5\) This case demonstrates a rare tumour at an unusual site and shows that aggressive surgical intervention can alter the outcome of an otherwise poor prognosis.

Fig. 1. Picture of resected specimen demonstrating intimate relationship to superior mesenteric vein. [wire passed through resected segment of vein].

References


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