Case report

Regional pancreatectomy and transverse colectomy with mesenteric vascular reconstruction for inflammatory pseudotumour of the head of pancreas and mesenteric root

DG Lall[1], ND Karanjia[1] and AEB Giddings[2]

[1]Royal Surrey County Hospital, Guilford, Surrey and [2]St. Thomas' Hospital, London, UK.

Background

Inflammatory myofibroblastic pseudotumour is a rare pancreatic lesion.

Case Outline

A 32-year-old woman with such a tumour was treated by a radical operation comprising proximal pancreaticduodenectomy (Whipple Procedure) and transverse colectomy with resection and reconstruction of the superior mesenteric artery and vein. She remains well 6 years later.

Discussion

The importance of aggressive surgical clearance rather than chemotherapy is highlighted in the management of patients with these unusual tumours.

Key words

Inflammatory myofibroblastic pseudotumour, fibrosarcoma, resection

Introduction

Inflammatory myofibroblastic tumour is a rare pancreatic lesion that is difficult to distinguish from fibrosarcoma. We present a case of such a tumour in a young woman that requires a very radical excision to prevent the risk of local recurrence.

Case Report

A 32–year-old woman was admitted with a 3-month history of upper abdominal discomfort, vomiting and weight loss. Abdominal examination suggested an epigastric mass, and a succussion splash was audible. A contrast study showed obstruction of the pyloric channel, and a CT scan showed a large cystic mass in the head of pancreas. At laparotomy the mass was found to involve the head, neck and body of the pancreas. It involved the pylorus and duodenum and entered the root of the transverse mesocolon, encasing the proximal transverse colon. In addition, the superior mesenteric artery (SMA) and vein (SMV) lay in the centre of the tumour with no plane of dissection (Figure 1). A frozen section showed that this was a spindle cell tumour of low grade malignancy. The presumptive diagnosis was that of a low-grade sarccoma.

Resection was possible only by sacrificing the superior mesenteric artery and vein. An extended pancreatoduodenectomy was carried out taking a segment of the SMA from its origin, up to and including the middle colic artery. In addition a segment of portal vein and SMV including the confluence with the splenic vein was taken. A transverse colectomy was performed en-bloc.

The splenic artery was divided along the upper border of the tail of pancreas. The vessel was mobilised and anastomosed to the SMA. The SMV and the stump of the portal vein above the tumour were mobilised, and an end-to-end anastomosis was constructed. Thus the small intestine was successfully revascularised (Figure 2). A standard Kausch-Whipple reconstruction was carried out with an end-to-end pancreatojejunostomy, an end-to-side hepatodochojejunostomy and an end-to-side valved gastrojejunostomy. The hepatic and splenic flexures were mobilised and anastomosed. She made an uncomplicated recovery.

The operative specimen showed severe compression of the mesenteric vessels, with the lumen of the SMV reduced to a tiny slit. The histological appearances (assessed locally and at the Royal Marsden Hospital) were said to lie between inflammatory myofibroblastic tumour and a very

Correspondence to: *Mr N Karanjia, Royal Surrey County Hospital, Egerton Road, Guildford, Surrey GU2 5XX, UK (e-mail: nkaranjia@aol.com)*

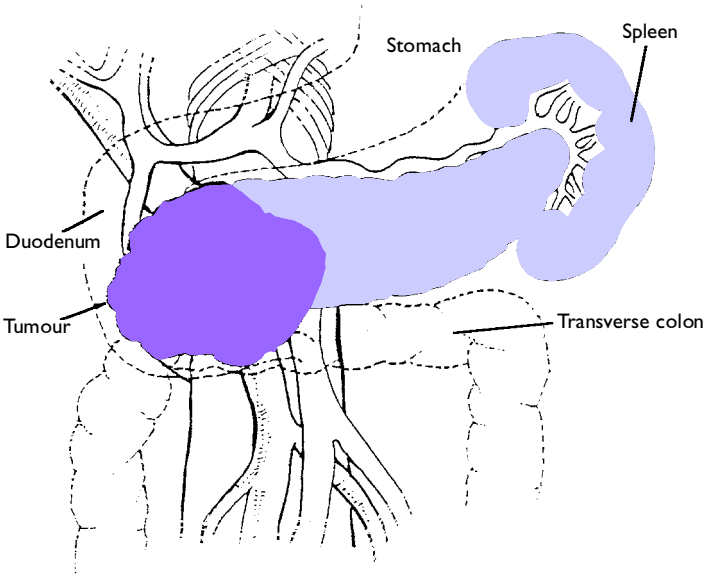

Figure 1. *Schematic diagram showing the position of the tumour in the pancreatic head.*

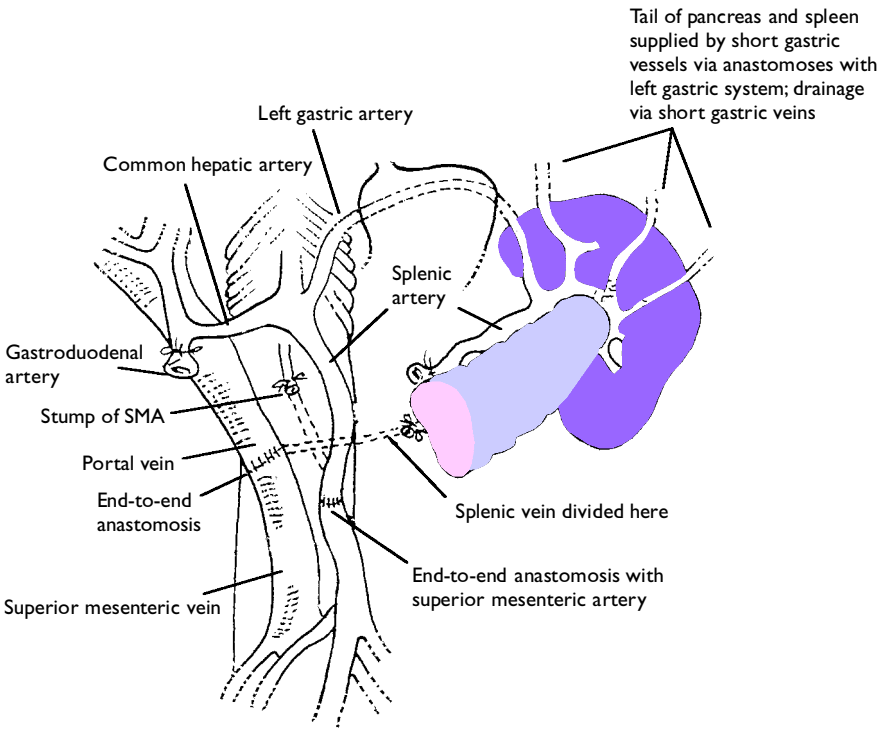

Figure 2. *Schematic diagram showing vascular anatomy and vascular reconstruction after resection of the tumour by Whipple's procedure with en-bloc transverse colectomy and excision of both superior mesenteric artery and superior mesenteric vein.*

low grade of fibrosarcoma. The resection margins were all microscopically clear of disease.

Initial postoperative problems included weight loss and diarrhoea. These were due to malabsorption from pancreatic insufficiency and rapid gut transit owing to denervation of the small intestine, as the nerve plexuses that travel with the mesenteric vessels were sacrificed. Her gut transit time measured 3 months postoperatively was 30 minutes. Preservation of the spleen and tail of pancreas has left her normoglycaemic, she has required high-dose supplementation with pancreatic extracts.

One year after operation she remained well and was gaining weight. Left costal pain proved to be a problem and CT scanning suggested disease recurrence. Upper gastrointestinal endoscopy demonstrated a normal stomach remnant without gastric varices, and a colonoscopy was unremarkable. She was subsequently re-explored, found to be free of recurrence and remains well nearly 6 years later.

Discussion

Inflammatory myofibroblastic tumour and low grade inflammatory fibrosarcoma form a histological and clinical spectrum of disease [1–3] separated by degree rather than an absolute difference. Some authors assert that the former tumour is benign [3], probably a post-inflammatory process and one that predominantly affects children, with a predisposition towards the lung [3]. In contrast, others have described a course indistinguishable from that of a malignant neoplasm, with a tendency to local recurrence, rapid growth and local tissue infiltration; indeed, there are reports in the literature of patients who have developed metastatic disease [2,4]. These tumours also affect adults, with a predisposition to the head and neck as well as intra-abdominal sites [4].

Histological variability of the disease process is reflected in the various terms used in its description: 'inflammatory pseudotumour', 'inflammatory myofibroblastic tumour' and even 'inflammatory fibrosarcoma' have all been applied by different authors [3,4]. It is agreed that the myofibroblast is the predominant cell type in these tumours, lying within a variable stroma interspersed with plasma cells, both B and T lymphocytes and mast cells [3,4]. The absence of nuclear pleomorphism, abnormal mitoses or hyperchromasia tends to point towards a benign rather than a malignant disease process.

Clinical presentation is generally with the symptoms and signs attributable to a mass lesion; the patient described here presented with gastric outflow obstruction and abdominal pain. There are reports of patients with inflammatory myofibroblastic tumour presenting with symptoms suggestive of a paraneoplastic syndrome [5], i.e pyrexia of unknown origin, raised erythrocyte sedimentation rate (ESR), a microcytic hypochromic anaemia refractory to iron, thrombocytosis, hypoalbuminaemia and a polyclonal hypergammaglobulinaemia, as well as local symptoms and signs of a mass lesion. In these patients surgical exploration demonstrated a single tumour mass and their symptoms resolved after resection. Furthermore, tumour recurrence was heralded by a return of these non-specific symptoms and signs, which again resolved on re-excision [5].

Anatomically there are some important aspects. As the splenic artery was diverted to supply the small intestine, the tail of pancreas and the spleen are now supplied by reversed flow through the short gastric arteries via their anastomoses with the left gastric system in the body of the stomach. Both organs appear to be adequately supplied and at the second look laparotomy a year later the spleen was normal in size. Isolated division of the splenic vein with an intact splenic artery could lead to left-sided portal (sectorial) hypertension with the development of gastric varices. However, in this case division of the splenic artery has probably reduced the pressure in the system, and therefore the risk of gastric varices is likely to be reduced.

Surgical operation is the treatment of choice in patients with inflammatory myofibroblastic pseudotumour; complete resection offers a good prognosis and likelihood of cure [2,3]. Accurate tissue diagnosis is paramount, for if the tumour is misdiagnosed as a soft tissue sarcoma palliative surgery only may be performed, or chemo-radiotherapy may be given as an adjunct. Both of these measures have been tried, but small numbers of patients and the unpredictable behaviour of the disease mean that it is impossible to draw meaningful conclusions as to their effectiveness.

Although the behaviour of these tumours is unpredictable, the most likely problem is local recurrence following an inadequate resection [2,3] or seeding of tumour if the capsule has been breached intra-operatively. Recurrences should be aggressively sought and, if amenable to operation, repeat resections should be performed. As the time-course for recurrence is unpredictable [2,3] prolonged follow-up is necessary, with appropriate imaging or even surgical re-exploration if indicated to ensure clearance of tumour.

The patient described above presents an exceptionally unusual pathological problem correctly managed by regional pancreatectomy, transverse colectomy and vascular reconstruction, which has provided the patient with her only chance of cure.

References

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