Reminder of important clinical lesson

Small bowel liposarcoma—a rare cause of small bowel perforation

K S Benaragama,¹ S K Neequaye,² D Maudgil,² A G Gordon²¹ Department of General Surgery, Heatherwood and Wexham Park Hospitals NHS Trust, Wexham Park Hospital, Slough, UK² Department of Radiology, Wexham Park Hospital, Slough, UK

Correspondence to K S Benaragama, shanka@doctors.org.uk

Summary

A 76-year-old patient was admitted with right-sided lower abdominal pain. CT scan of the abdomen demonstrated a fatty mass arising around the bowel in the left iliac fossa causing local distortion of the vasculature.

His clinical condition rapidly deteriorated with worsening abdominal signs necessitating an exploratory laparotomy. A segment of perforated small bowel associated with mesenteric mass was excised and the patient died 3 days later.

The histology demonstrated a well-differentiated liposarcoma. Liposarcomas are the most common soft tissue sarcomas in adults but occurrence in the gastrointestinal tract is extremely rare. The clinical presentation of these tumours is frequently non-specific and commonest symptoms are generalised, intermittent abdominal pain.

Surgical resection with clear margins is the treatment of choice for primary liposarcomas. They are moderately radiosensitive and chemotherapy is non-effective.

Although gastrointestinal liposarcomas have been previously reported, this is the first reported case of a primary liposarcoma associated with a small bowel perforation.

BACKGROUND

The case reported here is the first case of a primary liposarcoma of the small bowel presenting with spontaneous perforation of the bowel.

CASE PRESENTATION

A 76-year-old man was admitted with a 2-day history of acute onset right-sided lower abdominal pain. It was a constant and gradually worsening type of pain. There was no history of nausea, vomiting, fever, loss of appetite or loss of weight, or disturbance of bowel function.

He had a background history of left-sided pleural effusion, chronic renal failure, hypertension, chronic anaemia and two recent cerebrovascular events.

On examination he was haemodynamically stable with mild pallor (haemoglobin 10.1 g/dl, mean corpuscular volume 83, mean corpuscular haemoglobin concentration 313). He was mildly tender in the right iliac fossa with no signs of localised peritonism or palpable masses. On auscultation, the bowel sounds were present with normal intensity. The inflammatory markers (C reactive protein) were normal with slightly raised leucocytes (13.4×10^9). During the first 24 h in hospital, his haemoglobin level dropped further to 7 g/dl requiring blood transfusion.

INVESTIGATIONS

Plain abdominal x-ray and erect chest x-ray were unremarkable. Given his background of anaemia and right-sided abdominal pain, contrast enhanced CT scan showed the appearances of a possible internal hernia/volvulus or small bowel tumour. CT scan was performed because the patient was not peritonitic and had major morbidity, which made

laparotomy very high risk. This demonstrated a fatty mass arising around the bowel in the left iliac fossa causing local distortion of the vasculature. Emergency mesenteric angiography was arranged to investigate the rapidly falling haemoglobin and the plan was to attempt embolisation of the presumed bleeding point to avoid the high-risk laparotomy. This revealed considerable architectural distortion of ileal arcades with normal vascularity (figures 1 and 2).

DIFFERENTIAL DIAGNOSIS

Perforated appendix, acute gastrointestinal bleeding and intestinal obstruction.

TREATMENT

Twenty-four hours since the angiographic embolisation, an emergency exploratory laparotomy was performed. A segment of perforated small bowel (ileum) associated with a 20 cm mesenteric mass was excised. There was bleeding into the small bowel, but the tumour had perforated and there was diffuse fatty tumour material within the peritoneal cavity and a large mass, which was the liposarcoma that had burst its capsule.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient was managed in the intensive care unit where he died 3 days later of multi-organ failure, principally respiratory failure.

The histological sections show small bowel with an attached encapsulated cellular tumour composed of adipocytes of different sizes with several blood vessels and scattered inflammatory cells. Although classical lipoblasts are not seen, there were enlarged, slightly hyperchromatic

Figure 1 Axial CT scan demonstrating fatty mass arising around the bowel in left iliac fossa, causing local distortion of vasculature (arrow indicates the tumour).

nuclei within the tumour. The adipocytes were stained with Oil Red O and, immunohistochemically, the tumour cells/lipoblasts were positive for S-100 protein.

The histology of the small bowel lesion demonstrated a well-differentiated liposarcoma (table 1).

Table 1 Histopathology of the small bowel lesion

Macroscopy	A small bowel segment measuring 250 mm in length connected to a cystic structure contains whitish yellow friable material and blood clots and is covered by fibrinopurulent exudates probably tumour tissue attached to the inner aspect of the wall
Microscopy	The sections show small bowel with an attached encapsulated cellular tumour composed of adipocytes of different sizes with several blood vessels and scattered inflammatory cells. Although classical lipoblasts are not seen, there were enlarged, slightly hyperchromatic nuclei within the tumour. The adipocytes were stained with Oil Red O and immunohistochemically, the tumour cells/lipoblasts were positive for S-100 protein. The features are compatible with well-differentiated liposarcoma

DISCUSSION

Liposarcomas are the most common soft tissue sarcomas in adults accounting for approximately 16–18% of all soft tissue neoplasms.¹

Liposarcomas occurring in the gastrointestinal tract are extremely rare. They are among the least common malignant tumours and have been reported to occur in the stomach, jejunum, ileo-caecal valve and retro-peritoneum. Most of these tumours arise de novo and are believed to take origin from the undifferentiated perivascular mesenchyme.²

The clinical presentation of these tumours is frequently non-specific. The commonest symptoms are generalised, intermittent abdominal pain and bleeding usually as frank haemorrhage, melaena or occult blood in the faeces.³

WHO classifies these tumours into four categories: well differentiated, myxoid/round cell, pleomorphic and dedif-

ferentiated liposarcomas.⁴ The myxoid and the well-differentiated types are the commonest and the latter has the best prognosis; the 5-year survival rate ranges between 75% and 100%.⁵ The myxoid liposarcomas with associated round cell component are said to have a substantially higher risk of metastasis.⁶ Overall metastatic rates for liposarcoma have been reported to range from 37–50%.⁷

Surgical resection with clear margins whenever possible is the treatment of choice for primary liposarcomas.^{8,9} They are moderately radiosensitive with no evidence that chemotherapy is effective. Neither treatment has been proven to be of benefit in increasing long-term survival¹⁰; however, this is such a rare tumour that it is difficult to accurately estimate treatment effect.

There is only a handful of literature published on liposarcomas associated with intestinal perforation. In 1984, the

Figure 2 Hypercellular area with possible lipoblasts, cell size variation and focal hypercellularity.

title of Orzechowski and Wlodarczyk's paper in the Medline database indicated that liposarcoma of the intestinal mesentery caused peritonitis. The article was written in Polish and no abstract available. Horiguchi *et al* described a case of liposarcoma with peritonitis due to jejunal perforation. It reported the suspected cause of the perforation was adhesions due to a previous operation and was not due to direct infiltration of tumour cells because tumour cells were not observed at the perforation site.¹¹

We believe this is the first reported case of primary liposarcoma associated with a small bowel perforation (confirmed with the histological findings) and represents an important learning point. In retrospect, this patient was clinically very high-risk for anaesthesia and was septic with rapidly falling haemoglobin. Also the presence of a mesenteric mass with distortion of the vasculature caused some uncertainty leading to further investigation.

If not for the above reasons, an earlier laparotomy may have been associated with a more favourable outcome.

Learning points

- ▶ Although, the liposarcomas occurring in the gastrointestinal tract are extremely rare they can cause intestinal perforation.
- ▶ The clinical presentation of these tumours is frequently non-specific.
- ▶ This unique case represents an important learning point for all clinicians and pathologists, particularly in this age of increased access to cross-sectional imaging. The simple lesson here is to treat the patient not the scan.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Kransdorf MJ.** Malignant soft-tissue tumours in a large referral population: distribution of diagnoses by age, sex and location. *AJR Am J Roentgenol* 1995;**164**:129–34.
2. **Mohandas D,** Chandra RS, Srinivasan V, *et al.* Liposarcoma of the ileum with secondaries in the liver. *Am J Gastroenterol* 1972;**58**:172–6.
3. **Wilson JM,** Melvin DB, Gray GF, *et al.* Primary malignancies of the small bowel: a report of 96 cases and review of the literature. *Ann Surg* 1974;**180**:175–9.
4. **Christopher DM,** Unni KK, Mertens F. *WHO classification of tumours. Pathology and genetics of tumours of soft tissue and bone.* Lyon, France: IARC Press, 2002.
5. **Kindblom LG,** Angervall L, Svendsen P. Liposarcoma a clinicopathologic, radiographic and prognostic study. *Acta Pathol Microbiol Scand Suppl* 1975:1–71.
6. **Spillane AJ,** Fisher C, Thomas JM. Myxoid liposarcoma—the frequency and the natural history of nonpulmonary soft tissue metastases. *Ann Surg Oncol* 1999;**6**:389–94.
7. **Weiss SW,** Goldblum JR. Liposarcoma. In: Enzinger F, Weiss S, eds. *Enzinger and Weiss's soft tissue tumours.* 4th edn. St Louis, Missouri, USA: Mosby, 2001:641–93.
8. **Sato T,** Nishimura G, Nonomura A, *et al.* Intra-abdominal and retroperitoneal liposarcomas. *Int Surg* 1999;**84**:163–7.
9. **Fernández-Trigo V,** Sugarbaker PH. Sarcomas involving the abdominal and pelvic cavity. *Tumori* 1993;**79**:77–91.
10. **Moyana TN.** Primary mesenteric liposarcoma. *Am J Gastroenterol* 1988;**83**:89–92.
11. **Horiguchi H,** Matsui M, Yamamoto T, *et al.* A case of liposarcoma with peritonitis due to jejunal perforation. *Sarcoma* 2003;**7**:29–33.

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