CASE REPORT

Gastrointestinal stromal tumour (GIST) presenting as a strangulated inguinal hernia with small bowel obstruction

Marco Massani,¹ Giovanni Capovilla,² Cesare Ruffolo,¹ Nicolò Bassi^{1,2}

¹Regional Center for HPB Surgery, Regional Hospital of Treviso, Treviso, Italy

²Department of Surgical Oncological and Gastroenterological Sciences, University of Padova, Padova, Italy

Correspondence to

Dr Giovanni Capovilla, giovannicapovilla88@gmail.com

Accepted 5 January 2017

SUMMARY

Gastrointestinal stromal tumours (GISTs) can arise everywhere along the gastrointestinal (GI) tract. Their presentation in unusual locations should always be taken into account. A 74-year-old man referred to the emergency department for small bowel obstruction caused by an incarcerated inguinal hernia. A CT scan showed a mesenchymal tumour originating from the herniated bowel loop and a mass in the ascending colon. Laparoscopic resection of the mass and laparoscopic right hemicolectomy were performed. The histology showed a ruptured GIST arising from the herniated small bowel and a high-grade dysplasia villous adenoma of the right colon. GISTs can present with symptoms spanning from vague abdominal discomfort to surgical urgencies. Strangulated hernia is an extremely rare presentation, with only two cases described in the literature. A safe surgical approach was obtained with laparoscopy, maintaining surgical radicality. The ileal localisation and the pseudocapsule rupture were the main risk factors on prognostic stratification.

BACKGROUND

Gastrointestinal stromal tumours (GISTs) are rare mesenchymal tumours believed to arise from the interstitial cells of Cajal (ICC). These cells are usually referred to as the pacemaker of the gastrointestinal (GI) tract, forming the interface between the autonomic innervation of the bowel wall and the smooth muscle.¹

Although GISTs are rare tumours, they represent the most frequent mesenchymal tumour of the GI tract, accounting for 1–3% of all GI malignancies.^{1–4} The annual incidence is estimated to be around 7–20 per million population per year;^{1 5–7} however, recent data suggest that a much higher number of microscopic, subclinical lesions could be found; in particular, the frequency of subcentimetric gastric GISTs could be higher than previously estimated.^{8 9}

The median age at presentation is around 60–65 years; however, GISTs can arise at any age, with a similar frequency reported in men and women.^{10–12}

Around 95% of GISTs arising in adults express the product of the KIT proto-oncogene CD117 that is part of the KIT transmembrane receptor tyrosine kinase (RTK). Approximately 80% of these tumours harbour KIT gene mutations leading to constitutive activation of the KIT receptor that ultimately leads to tumour growth. Another 5–10%

of GISTs are KIT negative and have a mutation in the gene encoding the related PDGFRA RTK.^{1 13–15}

GISTs can arise in any portion of the GI tract; the most common primary sites are the stomach (60%) and small intestine (35%). Less common primary sites include the duodenum (4–5%) and rectum (4%). Only a few cases have been reported in the oesophagus (<1%), colon and appendix (1%).¹⁶ Clinical presentation is strictly dependent on the site and size of the tumour; overall, the most frequent symptoms are GI bleeding, abdominal pain, anaemia and abdominal mass.

In the majority of cases, GISTs occur as single lesions. Association with other tumours is rare and usually typical of several autosomal dominant familial syndromes (neurofibromatosis type 1, Carney triad and familial GIST).^{17–19}

Imaging has a fundamental role in the clinical diagnosis of GISTs, as a tumour biopsy carries the risk of rupture or seeding in the biopsy tract.²⁰ Endoscopic ultrasound (EUS) and CT are the most commonly used imaging tools.

We present the case of a 74-year-old man who referred to our institution for bowel obstruction due to an incarcerated inguinal hernia with an incidental finding of a small bowel GIST located in the hernia sac and a synchronous high-grade dysplasia villous adenoma of the right colon.

CASE PRESENTATION

Patient

A 74-year-old man presented to our emergency department with a history of 2 days of diffuse abdominal pain associated with nausea, vomiting and the presence of a mass in the right groin. There were no other GI or genitourinary symptoms, and the patient had an unremarkable medical history with no prior abdominal surgery.

On physical examination, the abdomen appeared soft but diffusely distended, with tinkling bowel sounds. A tender, reducible, non-pulsatile mass was revealed in the right groin. The blood investigation results were unremarkable. A simple abdominal X-ray was performed showing multiple dilated loops of a gas-filled bowel.

The patient was initially managed with manual reduction of the inguinal hernia, intravenous fluid resuscitation, nasogastric tube insertion and catheterisation. CT imaging with intravenous contrast was performed revealing a 3 cm expansive mass in the ascending colon and the right inguinal herniation of a distal ileal loop that showed reduced

CrossMark

To cite: Massani M, Capovilla G, Ruffolo C, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-217273

parietal enhancement coherent with an ischaemic process (figure 1). The proximal portion of the herniated loop presented a vascularised, intraparietal, oval-shaped lesion, which measured 6 cm in diameter. Under laparoscopic surgical exploration, the presence of distended bowel loops sustained by an incarcerated inguinal hernia was confirmed. After reduction of the herniated loop, a solid exophytic mass protruding from the distal ileum was revealed (figure 2). The lesion was located about 60 cm from the ileocecal valve and entered the inguinal canal through the internal inguinal orifice causing the kinking of the adjacent bowel loop. After accurate exploration of the abdominal cavity confirming the absence of secondary lesions, a laparoscopic right hemicolectomy was performed with extracorporeal side-to-side manual ileocolonic anastomosis through a right pararectal incision. The same access was used to perform a local resection of the ileal lesion with an intestinal stapler, leaving no residual macroscopic disease. Prosthetic repair of the inguinal canal was ultimately performed through right inguinoscopy. Postoperatively, the patient recovered and was discharged on day 7 after the procedure without complications.

Pathologic findings

On gross examination, the ileal lesion appeared as a soft polypoid mass measuring about 7 cm in greater diameter. The mass presented necrosis and suffusion haemorrhage on the surface.

The pathology report was GIST with spindle cells morphology, apparently unaffected surgical margins (R0) and a mitosis index of 2/5 mm³. The immunohistochemistry was positive for CD117 (c-kit) and DOG1 (figure 3) and negative for CD34 and S100. Mutational analysis revealed the presence of a c-kit exon 11 mutation c.1676T>A (p.Val559Asp). The risk of progression according to the Miettinen-Lasota criteria would have been moderate (24%); however, tumour rupture was observed on microscopy, thus establishing a high risk of disease progression.^{4 16 21} On opening the ascending colon specimen, a sessile mass measuring 4.5×3 cm was revealed. Histology showed evidence of a villous adenoma with high-grade dysplasia. Fourteen lymph nodes were collected with no histological sign of metastatisation.

DIFFERENTIAL DIAGNOSIS

The pathological diagnosis of GISTs relies on the demonstration of CD117 (c-kit) and DOG1 expression by immunohistochemistry.²² The expression of CD117 and DOG1 is extremely useful in differentiating GISTs from other soft tissue malignancies with a spindled appearance at histology that are CD117/

DOG1 negative, such as GI leiomyosarcoma, malignant fibrous histiocytoma and dedifferentiated liposarcoma.²³

DISCUSSION

Only 70% of patients with GISTs are symptomatic, while 20% of the lesions are characterised by the absence of a clinical picture and 10% are incidentally detected on autopsy. The clinical presentation of small bowel GISTs can be extremely variable according to their site and size spanning from nausea and vague abdominal discomfort to acute abdominal pain, GI bleeding and small bowel obstruction (SBO).²⁴⁻²⁷ In a large series of 906

Figure 2 Surgical specimen: a solid exophytic mass protruding from the distal ileum was located about 60 cm from the ileocecal valve and entered the inguinal canal through the internal inguinal orifice causing the kinking of the adjacent bowel loop. Signs of haemorrhagic necrosis can be seen on the surface of the lesion.

Figure 1 Computerised tomographic imaging in the portal phase showing the right inguinal herniation of a distal ileal loop with reduced parietal enhancement coherent with an ischaemic process and the presence of an intraparietal, oval-shaped, contrast-enhanced mass.

Figure 3 (A, B) Pathology showing atypical spindle cells on the H&E stain at (A) ×20 and (B) ×40. (C) Immunohistochemistry showing CD117 expression (×40). (D) Immunohistochemistry showing DOG1 expression (×40).

patients published by Miettinen *et al*,²⁸ jejunal and ileal GISTs most commonly presented with GI bleeding or an acute abdomen. In a recent paper by Xing *et al*,²⁹ GIST symptoms at presentation were evaluated in 197 patients on the basis of their location in the small bowel. Cardinal symptoms for patients with ileal GISTs included epigastric discomfort (38.3%), abdominal mass (34%), incidental finding (27.7%) and GI bleeding (12.8%). Conversely, patients with duodenal GISTs most frequently presented with GI bleeding (25%), followed by incidental finding (22.1%), epigastric discomfort (19.1%) and abdominal mass (7.1%).

In a recent series of 92 patients admitted with different emergency presentations of GISTs, Sorour *et al*³⁰ reported GI bleeding (48.9%) and SBO (28.3%) to be the most frequent presenting symptoms. Of 27 patients with small bowel GISTs, 22 showed a picture of SBO in the form of intussusception (12 cases) or lumen obstruction due to exophytic and endophytic growth (10 cases).

In our case, the clinical picture of SBO was probably due to the exophytic lesion entering the internal inguinal orifice, stretching the adjacent bowel loop and ultimately causing its kinking. It is not easy to ascertain whether this was an acute event in the context of a pre-existing inguinal hernia or the result of a chronic process involving the growth of the lesion in the inguinal canal; in our opinion, the former hypothesis seems to be more probable, considering the paucity of adhesions and the relatively easy reduction of the herniated content from the inguinal canal.

Extremely rare sites of GIST localisation have been reported; remarkable examples are Meckel's diverticulum and vermiform appendix, the latter with <20 cases described in the literature.^{31–34}

To the best of our knowledge, this is the third reported case of a GIST presenting as an inguinal hernia. In the case published by Mulla *et al*,³⁵ the patient developed a large, irreducible right inguinal mass while on the waiting list for a bilateral inguinal hernia repair, showing no major symptoms. The patient underwent a CT scan showing a large abdominal mass with areas of necrosis and invasion into the caecum. An open wedge biopsy was then performed and, after confirmation of the diagnosis, the patient underwent chemotherapy with imatinib. In the case published by Tinoco-González,³⁶ the patient arrived in the emergency department reporting the sudden appearance of a mass in the left inguinal region. Suspecting the presence of an incarcerated inguinal hernia, urgent exploration of the inguinal canal was performed, revealing a jejunal loop containing a tumour mass. On conversion to an infraumbilical laparotomy, a segmental bowel resection was performed leaving macroscopic disease in the pelvis and the bladder. The author stated that a preoperative CT imaging would not have oriented towards the diagnosis given the rarity of the clinical picture.

In our institution, a CT scan of the abdomen and pelvis is routinely used in patients presenting with SBO since it can provide incremental information in an aetiology over a simple X-ray, eventually leading to changes in the management.^{37–38} The usefulness of CT varies for GIST according to tumour location. Its role is fundamental in the diagnosis of GISTs affecting the gut district, where traditional endoscopy is not accessible and percutaneous biopsy is contraindicated due to the risk of tumour dissemination.^{39–41} CT features offer valuable information in the differential diagnosis between mesenchymal tumours and lymphoma or epithelial neoplasms affecting the GI tract; however, further differentiation frequently necessitates histological assessment.

Small lesions often appear as sharply margined, smooth walled, homogeneous, soft tissue masses with moderate contrast enhancement. Large tumours show key features such as heterogeneous enhancement, central necrosis and cavitation.^{39–41} In the presented case, a CT scan was able to detect the presence of an oval-shaped, contrast-enhancing, homogeneous mass. This finding inevitably influenced the surgical decision, prompting a more cautious laparoscopic approach rather than the immediate repair of the inguinal hernia, with potential mishandling of the already ruptured lesion.

Indeed, laparoscopy is currently recommended for non-large GISTs in favourable anatomic locations such as the anterior wall of the stomach, jejunum and ileum.⁴² Small series and retrospective analyses demonstrated low recurrence rates, a shorter hospital stay and low morbidity with a laparoscopic approach compared to laparotomy.^{27–43–46} In a recent meta-analysis of 19 studies, Chen *et al*⁴⁷ concluded that laparoscopy is associated with less blood loss, lower complication rates and a shorter hospital stay, with no difference in long-term outcomes. In the presented case, laparoscopy allowed us to perform a safe resection of the ileal mass leaving no macroscopically affected margins. Despite the cautious handling of the tumour, the pseudocapsule appeared ruptured on microscopy; this finding was coherent with the intraoperative observation of suffusion haemorrhage on the tumour's surface. That considered, we feel that tumour rupture was the consequence of the prolonged ischaemia caused by the strangulation of the herniated mass in the inguinal canal rather than a consequence of the surgical manoeuvres.

The main prognostic factors for GISTs are mitotic rate, tumour size and tumour location. These criteria have been incorporated in a risk classification proposed by the Armed force Institute of Pathology (AFIP) which is still the most widely used in clinical practice.¹⁶ Although not included in the aforementioned risk classification, tumour rupture in the abdominal cavity, whether spontaneous or following surgical trauma, proved to harbour a risk of recurrence of nearly 100%.⁴⁸ In the light of these findings, a different risk stratification, including tumour rupture as a prognostic factor, was proposed by Joensuu⁴⁹ as a modification of the previous National Institute of Health (NIH) consensus stratification proposed by Fletcher *et al* (modified NIH).¹ More recently, prognostic contour maps were generated, which incorporate the mitotic index and tumour size as continuous non-linear variables, while tumour rupture is considered in addition to the tumour site.⁵⁰ A nomogram yielding the 10-year overall survival probability has also been developed treating tumour location as a categorical variable and tumour size and mitotic index as continuous variables and may represent an alternative, more practical approach in clinical decision-making.⁵¹

In the presented case, tumour size, non-gastric localisation and tumour rupture were the main risk factors. If we consider the AFIP classification, the tumour that we presented is classified as 'moderate risk' on measuring more than 5 cm, being localised in the small intestine and having a mitosis index of <5/5 mm³. The presence of a tumour rupture classifies the lesion as 'high risk' according to the modified NIH classification, irrespective of the size, localisation and mitotic index. Following both risk classification and given the presence of a favourable c-kit mutation (exon 11 mutation), the patient would benefit from adjuvant chemotherapy with imatinib.

The incidence of other primary malignancies in patients with GIST is usually typical of syndromic pictures originating from germ-line mutations in NF1, succinate dehydrogenase complex or KIT. However, the occurrence of other cancers in patients

with sporadic GISTs is a relatively frequent event. The reported incidence of additional primary tumours among patients with GIST varies from 2.95% to 33% in different studies; colorectal and gastric tumours seem to be the most common other primary neoplasms.⁵² Breast, prostate and renal cell carcinomas are also common.^{53–56} In a clinicopathologic study of 260 patients, Hechtman *et al*⁵⁶ reported an association between the increased mitotic rate in GIST ($\geq 5/50$ HPF) and the incidence of secondary malignancies of any chronology, while Kalmar discovered a greater incidence of synchronous or metachronous GI tumours in patients with GIST than in the general population.⁵⁷ However, at present, data supporting a potential correlation between GIST and other neoplasms are insufficient.

Learning points

- ▶ Gastrointestinal stromal tumour can present with a broad spectrum of symptoms spanning from vague symptoms to surgical urgencies.
- ▶ The localisation in unusual sites should always be taken into account.
- ▶ Gastrointestinal stromal tumour can present with a broad spectrum of symptoms spanning from vague symptoms to surgical urgencies.
- ▶ A CT scan is a reliable tool in the identification of mesenchymal tumours, even in the emergency setting.
- ▶ Laparoscopy, in experienced hands, allows the achievement of a surgically radical resection.
- ▶ Proper prognostic stratification is fundamental in order to offer the patient the proper post-surgical treatment.

Contributors MM and NB planned and designed the case report. GC collected and reviewed the literature on the subject and wrote the paper. MM performed the draft revision. CR collected the pathological and clinical data, provided the images and contributed with the other authors on the analysis and interpretation of data. NB provided the final revision and approval of the article.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Fletcher C, Berman JJ, Corless C, *et al*. Diagnosis of gastrointestinal stromal tumors: a consensus approach. *Human Pathol* 2002;33:459–65.
- 2 van der Zwan SM, DeMatteo RP. Gastrointestinal stromal tumor, 5 years later. *Cancer* 2005;104:1781–8.
- 3 Levy AD, Remotti HE, Thompson WM, *et al*. Gastrointestinal stromal tumors: radiologic feature with pathologic correlation. *Radiographics* 2003;23:283–304.
- 4 Casali PG, Dei Tos AP, Gronchi A. Gastrointestinal stromal tumors. In: De Vita VT, Hellman S, Rosenberg SA, eds. *Cancer: principles and practice of oncology*. Philadelphia (PA): Wolters Kluwer Health, 2015:745–54.
- 5 Tran T, Davila JA, El-Serag HB. The epidemiology of malignant gastrointestinal stromal tumors: an analysis of 1,458 cases from 1992 to 2000. *Am J Gastroenterol* 2005;100:162–8.
- 6 US population data from the US Census Bureau. 2008. <http://www.census.gov/popest/states/NST-ann-est.html> (accessed 8 Mar 2011).
- 7 Ma GL, Murphy JD, Martinez ME, *et al*. Epidemiology of gastrointestinal stromal tumors in the era of histology codes: result of a population-based study. *Cancer Epidemiol Biomarkers Prev* 2015;24:298–302.
- 8 Kawanowa K, Sakuma Y, Sakurai S, *et al*. High incidence of microscopic gastrointestinal stromal tumors in the stomach. *Hum Pathol* 2006;37:1527–35.
- 9 Agaimy A, Wunsch PH, Hofstaedter F, *et al*. Minute gastric sclerosing stromal tumors (GIST tumorlets) are common in adults and frequently show c-kit mutations. *Am J Pathol* 2007;31:113–20.
- 10 Besana-Ciani I, Boni L, Dionigi G, *et al*. Outcome and long term results of surgical resection for gastrointestinal stromal tumors (GIST). *Scand J Surg* 2003;92:195–9.
- 11 De Matteo RP, Lewis JJ, Leung D, *et al*. Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. *Ann Surg* 2000;231:51–8.
- 12 Badalamenti G, Rodolico V, Fulfaro F, *et al*. Gastrointestinal stromal tumors: focus on histopathological diagnosis and biomolecular features. *Ann Oncol* 2007;6:136–40.
- 13 Heinrich MC, Corless CL, Duensing A, *et al*. PDGFRA activating mutations in gastrointestinal stromal tumors. *Science* 2003;299:708–10.
- 14 Hirota S, Ohashi A, Nishida T, *et al*. Gain-of-function mutations of platelet-derived growth factor receptor α gene in gastrointestinal stromal tumors. *Gastroenterology* 2003;125:660–7.
- 15 Ricci R, Dei Tos AP, Rindi G. GISTogram: a graphic presentation of the growing GIST complexity. *Virchows Arch* 2013;463:481–7.
- 16 Miettinen M, Lasota J. Gastrointestinal stromal tumors: pathology and prognosis at different sites. *Semin Diagn Pathol* 2006;23:70–83.
- 17 Behranwala KA, Spalding D, Wotherspoon A, *et al*. Small bowel gastrointestinal stromal tumours and ampullary cancer in type 1 neurofibromatosis. *World J Surg Oncol* 2004;2:1.
- 18 Carney JA, Sheps SG, Go VL, *et al*. The triad of gastric leiomyosarcoma, functioning extra-adrenal paraganglioma and pulmonary chondroma. *N Engl J Med* 1977;296:1517–18.
- 19 O'Riain C, Corless CL, Heinrich MC, *et al*. Gastrointestinal stromal tumors. Insights from a new familial GIST kindred with unusual genetic and pathologic features. *Am J Surg Pathol* 2005;29:1680–3.
- 20 Connolly EM, Gaffney E, Reynolds JV. Gastrointestinal stromal tumors. *Br J Surg* 2003;90:1178–86.
- 21 Rubin BP, Blanke CD, Demetri GD, *et al*. Protocol for the examination of specimens from patients with gastrointestinal stromal tumors. *Arch Pathol Lab Med* 2010;134:165–70.
- 22 Espinosa I, Lee CH, Kim MK, *et al*. A novel monoclonal antibody against DOG1 is a sensitive and specific marker for gastrointestinal stromal tumors. *Am J Surg Pathol* 2008;32:210–18.
- 23 Stanford Medicine Surgical Pathology Criteria. <http://surgepathcriteria.stanford.edu/gitumors/gist-gastrointestinal-stromal-tumor/differential-diagnosis.html#t3> (accessed 22 Nov 2016).
- 24 Goettsch WG, Bos SD, Breekveldt-Postma N, *et al*. Incidence of gastrointestinal stromal tumours is underestimated: results of a nation-wide study. *Eur J Cancer* 2005;41:2868–72.
- 25 Miettinen M, Lasota J. Gastrointestinal stromal tumors (GISTs): definition, occurrence, pathology, differential diagnosis and molecular genetics. *Pol J Pathol* 2003;54:3–24.
- 26 Yan BM, Kaplan GG, Urbanski S, *et al*. Epidemiology of gastrointestinal stromal tumors in a defined Canadian Health Region: a population-based study. *Int J Surg Pathol* 2008;16:241–50.
- 27 Rammohan A, Sathyanesan J, Rajendran K, *et al*. A gist of gastrointestinal stromal tumors: a review. *World J Gastrointest Oncol* 2013;5:102–12.
- 28 Miettinen M, Makhoul H, Sobin LH, *et al*. Gastrointestinal stromal tumors of the Jejunum and Ileum: a clinicopathologic, immunohistochemical, and molecular genetic study of 906 cases before imatinib with long-term follow-up. *Am J Surg Pathol* 2006;30:477–89.
- 29 Xing GS, Wang S, Sun YM, *et al*. Small bowel stromal tumors: different clinicopathologic and computed tomography features in various anatomic sites. *PLoS One* 2015;10:e0144277.
- 30 Sorour MA, Kassem MI, Ghazal Ael-H, *et al*. Gastrointestinal stromal tumors (GIST) related emergencies. *Int J Surg* 2014;12:269–80.
- 31 Chandramohan K, Agarwal M, Gurjar G, *et al*. Gastrointestinal stromal tumour in Meckel's diverticulum. *World J Surg Oncol* 2007;5:50–4.
- 32 Lorusso R, Forte A, Urbano V, *et al*. Small bowel stromal tumors in a "meckelian" location. About a clinical observation. *Ann Ital Chir* 2003;74:707–11.
- 33 De la Morena LF, Fernández-Salazar L, Velayos B, *et al*. Meckel's diverticulum and gastrointestinal stromal tumor: an unusual association. *Gastroenterol Hepatol* 2007;30:534–7.
- 34 Chun JM, Lim KH. Gastrointestinal stromal tumor of the vermiform appendix mimicking Meckel's diverticulum; case report with literature review. *Int J Surg Case Rep* 2016;21:20–2.
- 35 Mulla MG, Yeung J, Reynolds JR. Gastrointestinal stromal tumor (GIST) mimicking an indirect inguinal hernia, a rare presentation. *Eur Surg* 2007;39:270–2.
- 36 Tinoco-González J. Gastrointestinal stromal tumor (GIST) presenting as a groin mass mimicking and incarcerated hernia. *Int J Surg Case Rep* 2015;6:166–8.
- 37 Maung AA, Johnson DC, Piper GL, *et al*. Evaluation and management of small-bowel obstruction: an eastern association for the surgery of trauma practice management guideline. *J Trauma Acute Care Surg* 2012;73:S362–9.
- 38 Daneshmand S, Hedley CG, Stain SC. The utility and reliability of computed tomography scan in the diagnosis of small bowel obstruction. *Am Surg* 1999;65:922–6.
- 39 Lau S, Tam KF, Kam CK, *et al*. Imaging of gastrointestinal stromal tumour (GIST). *Clin Radiol* 2004;59:487–98.
- 40 Scarpa M, Bertin M, Ruffolo C, *et al*. A systematic review on the clinical diagnosis of gastrointestinal stromal tumors. *J Surg Oncol* 2008;98:384–92.

- 41 Kochhar R, Manoharan P, Leahy M, *et al.* Imaging in gastrointestinal stromal tumours: current status and future directions. *Clin Radiol* 2010;65:584–92.
- 42 NCCN Clinical Practice Guidelines in Oncology. http://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf (accessed 12 Mar 2016). Version 2.2016.
- 43 Beham AW, Schaefer IM, Schüller P, *et al.* Gastrointestinal stromal tumors. *Int J Colorectal Dis* 2012;27:689–700.
- 44 Pidhorecky I, Cheney RT, Kraybill WG, *et al.* Gastrointestinal stromal tumors: current diagnosis, biologic behavior, and management. *Ann Surg Oncol* 2000;7:705–12.
- 45 Stamatakis M, Douzinas E, Stefanaki C, *et al.* Gastrointestinal stromal tumor. *World J Surg Oncol* 2009;7:61.
- 46 Demetri GD, von Mehren M, Antonescu CR, *et al.* NCCN Task Force report: update on the management of patients with gastrointestinal stromal tumors. *J Natl Compr Canc Netw* 2010;8:S1–S44.
- 47 Chen K, Zhou YC, Mou YP, *et al.* Systematic review and meta-analysis of safety and efficacy of laparoscopic resection for gastrointestinal stromal tumors of the stomach. *Surg Endosc* 2015;29:355–67.
- 48 Hohenberger P, Ronellenfitsch U, Oladeji O, *et al.* Pattern of recurrence in patients with ruptured primary gastrointestinal stromal tumour. *Br J Surg* 2010;97:1854–9.
- 49 Joensuu H. Risk stratification of patients diagnosed with gastrointestinal stromal tumor. *Hum Pathol* 2008;39:1411–19.
- 50 Joensuu H, Vehtari A, Riihimäki J, *et al.* Risk of recurrence of gastrointestinal stromal tumour after surgery: an analysis of pooled population-based cohorts. *Lancet Oncol* 2012;13:265–74.
- 51 Rossi S, Miceli R, Masserini L, *et al.* Natural History of Imatinib-naïve GISTs: a retrospective analysis of 929 cases with long-term follow-up and development of a survival nomogram based on mitotic index and size as continuous variables. *Am J Surg Pathol* 2011;11:1646–56.
- 52 Agaimy A, Wunsch PH, Sobin LH, *et al.* Occurrence of other malignancies in patients with gastrointestinal stromal tumors. *Semin Diagn Pathol* 2006;23:120–9.
- 53 Pandurengan RK, Dumont AG, Araujo DM, *et al.* Survival of patients with multiple primary malignancies: a study of 783 patients with gastrointestinal stromal tumor. *Ann Oncol* 2010;21:2107–11.
- 54 Ferreira SS, Werutsky G, Toneto MG, *et al.* Synchronous gastrointestinal stromal tumors (GIST) and other primary cancers: case series of a single institution experience. *Int J Surg* 2010;8:314–17.
- 55 Wronski M, Ziarkiewicz-Wroblewska B, Gornicka B, *et al.* Synchronous occurrence of gastrointestinal stromal tumors and other primary gastrointestinal neoplasms. *World J Gastroenterol* 2006;12:5360–2.
- 56 Hechtman JF, DeMatteo R, Nafa K, *et al.* Additional primary malignancies in patients with Gastrointestinal Stromal Tumor (GIST): a clinicopathologic study of 260 patients with molecular analysis and review of the literature. *Ann Surg Oncol* 2015;22:2633–9.
- 57 Kalmár K, Tornóczy T, Póto L, *et al.* Gastrointestinal stromal tumours in a single institute: is there an association to other gastrointestinal malignancies?. *Magy Seb* 2004;57:251–6.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow