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Case Report

Maternal GIST in twin pregnancy: Case report of a rare and complex management challenge

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Case report

A 42 year old woman (G5 P3 + 1 twin pregnancy) presented for routine antenatal morphology ultrasound scan at 20 weeks. Ultrasound also demonstrated an incidental, solitary, 10 cm mass in the left retroperitoneum. Fine needle cytology revealed indeterminate spindle cells. The retroperitoneal mass did not change on close ultrasound surveillance and gestation progressed normally. Elective Caesarian section was scheduled at term with further investigation of the retroperitoneal lesion planned for the postnatal period. Past medical history was unremarkable except for one episode of gastrointestinal haemorrhage (capsule endoscopy, upper GI endoscopy and colonoscopy all negative) requiring multiple blood transfusions four years prior to the pregnancy.

At 36 weeks the patient presented with spontaneous, painless haematochezia and severe anaemia (Hb 56 g/L reference range 110–114 g/L) in combination with fulminating pre-eclampsia. Healthy twins were delivered by emergency Caesarian section. After delivery active rectal bleeding continued requiring aggressive resuscitation and transfusion. Nuclear labelled red blood cell scan showed brisk, active bleeding in the midabdomen and blood pooling in jejunal loops. Angiography confirmed bleeding from a hypervascular left retroperitoneal mass fed by multiple left sided aortic branches. Extravasation into the duodenum was noted. Several attempts at angiographic embolization failed. Postangiogram computed tomography demonstrated an 11 cm, heterogeneous vascular lesion eroding into the small bowel. The abnormal mass displaced the psoas medially, the left kidney posteriorly and the third part of the duodenum anteriorly (Fig. 1).

Urgent exploratory laparotomy was undertaken because of ongoing haemodynamic instability. At the operation, small bowel loops were distended with sanguineous fluid but no free intraperitoneal blood was present. A 10 cm highly vascular cystic retroperitoneal mass was seen to emanate from the third part of the duodenum (Fig. 2). A complete resection of the lesion with histologically clear margins was performed with primary closure of the duodenal defect. No metastatic disease was evident at the operation or during subsequent CT staging investigations. The patient went on to make an uneventful recovery.

Pathology reported a 120 mm encapsulated GIST containing areas of haemorrhage. The spindle cell neoplasm arose from muscularis propria of the duodenum and contained prominent, dilated vascular channels with foci suspicious for vascular invasion. Surgical margins were clear of tumour. Mitotic index was low (1/40 high powered field). Immunohistochemical markers were positive for CD117 and DOG1 and negative for CD34, PDGFR, desmin and S100.

GIST is a gastrointestinal tract neoplasm derived from precursors of interstitial cells of Cajal (Tran et al., 2005; Goldblum, 2002). Although GIST is the most frequent mesenchymal tumour of the digestive tract, it is still uncommon with an incidence of 6.5–14.5 per million (Tran et al., 2005). 46% of GIST patients are female and median age at diagnosis is 55 to 65 years (Tran et al., 2005). In females within the reproductive ages the tumour is rare. Two thirds of GISTs are derived from the stomach, one quarter from the small bowel and the remainder from the colon, rectum and oesophagus (Folgado Folgado et al., 2008; Goldblum, 2002). Four cases of GIST in pregnancy have been reported and all involved either the stomach or the small bowel (Barry, 2009; Scherjon et al., 2009; Valente et al., 1996). Common presentations of GIST include haemorrhage, anaemia and abdominal mass although many are discovered incidentally (Goldblum, 2002). Previously GIST detected incidentally on antenatal ultrasound scan has been misdiagnosed as both ovarian and adnexal in pathology (Valente et al., 1996; Scherjon et al., 2009; Barry, 2009).

Biological behaviour of GIST ranges from indolent to aggressive and all GIST > 2 cm are considered to have malignant potential. Prognostication for primary tumours is based on molecular biomarkers,

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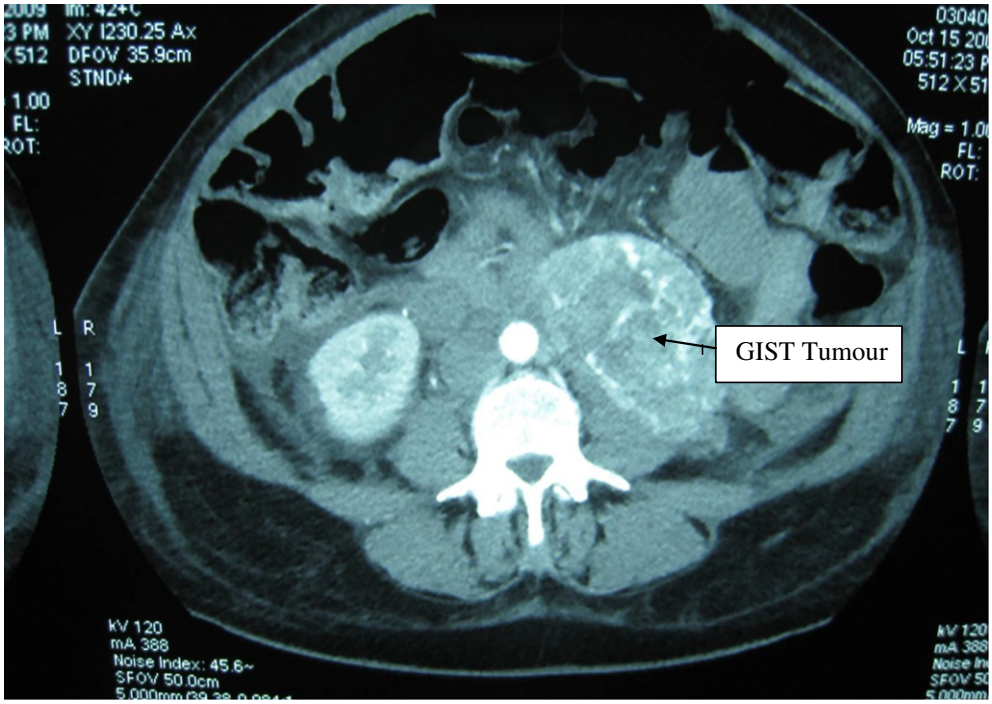

Fig. 1. A CT scan showing the GIST tumour displacing the kidney, the psoas muscle and the 3rd part of the duodenum.

mitotic index, tumour size, tumour location, resectability and the presence of metastasis (Tran et al., 2005; Folgado et al., 2008; Goldblum, 2002). Small bowel GISTs are considered to have the highest risk of metastatic progression (Goldblum, 2002). Most GISTs have activating mutations in KIT (75–80%) or PDGFRA (5–10%) (Folgado et al., 2008; Goldblum, 2002; Tran et al., 2005). Overall the 5 year survival rate for GIST patients is 42% (Tran et al., 2005). Of the four previously reported cases of GIST in pregnancy, three patients survived a minimum of nine years post diagnosis despite presenting with metastatic disease (Barry, 2009; Scherjon et al., 2009; Valente et al., 1996). The case presented here suggests that GIST had been in situ at least four years prior to diagnosis as obscure gastrointestinal haemorrhage was first investigated in the patient at that time.

GIST is predominantly managed with surgery and tyrosine kinase inhibitors as conventional chemotherapy and radiotherapy are ineffective. Surgery aims at complete tumour removal and is generally safe in pregnancy. Tyrosine kinase inhibitors (TKIs) such as imatinib mesylate are approved for use in recurrent, unresectable or metastatic GIST. TKI's have shown a marked improvement in survival (Perez et al., 2006). However imatinib and its metabolites occur in active concentrations in maternal blood, placenta and breast milk (Russel et al., 2007). There are reports of spontaneous abortions and birth defects following periconception or intrauterine exposure to imatinib (Ault et al., 2006). As a result TKIs are contra-indicated in pregnancy and breastfeeding (Wolf and Rumpold, 2009).

In patients with a history of GI bleeding and pelvic/abdominal mass, a GIST tumour should be considered in addition to other tumours of GI origin. Given the difficulties, pregnant patients with GIST are best managed by a multidisciplinary team with expertise in GI tumours and foetal–maternal medicine.

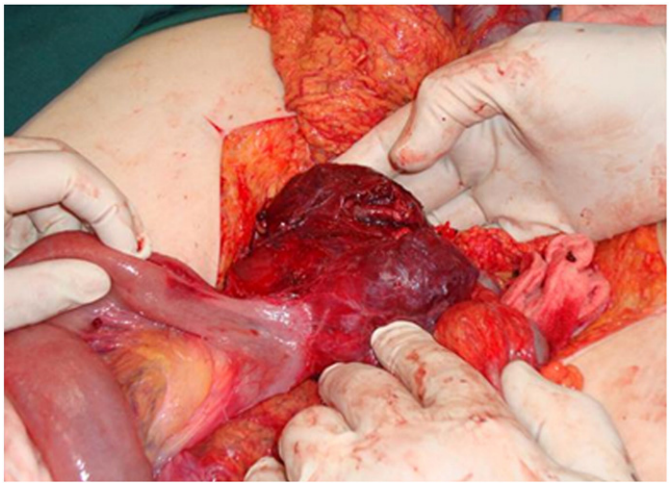

Fig. 2. An intra-operative photograph of the tumour.

Conflict of interest statement
The authors declare that there is no conflict of interest.

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