CASE REPORT

The mass effect of a slowly growing GIST

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SUMMARY

Gastrointestinal stromal tumours (GISTs) are typically defined as solid masses arising from the GI tract, most commonly from the stomach and small intestine. They seldom present in a cystic form. Management of cystic masses arising from the GI tract may pose a diagnostic predicament. We had one such case that presented itself with complaints of a slow growing intra-abdominal mass. An ultrasound scan demonstrated a thick-walled cystic lesion arising from the pelvis. Further imaging evaluations in the form of a CT scan revealed a complex large cystic mass arising from left upper quadrant (see Figure 1). Due to the uncertainty of origin of this mass and lack of invasion or lymphadenopathy, it was thought to be benign. After a multidisciplinary meeting, it was concluded that an urgent surgical excision of this benign mass was the best treatment. The surgical treatment of which entailed a 10 hours surgery to resect this 10 kg lesion, which comprised 7 L fluid and 3 kg solid mass. Histopathology aided in the diagnosis of this lesion as a CD117-positive and DOG1-positive GIST.

BACKGROUND

This case is of a 65-year-old man who presented with a slow-growing large intra-abdominal mass, which presented itself in an almost 'gravid' fashion. This mass displaced the pancreas into the pelvis and was diagnosed as a cystic gastrointestinal stromal tumour (GIST) intraoperatively and confirmed by immunochemistry and histopathology studies. The patient fully responded to the multimodal treatment given and despite a moderate to high recurrence prediction via the prognostic algorithm, there has been no recurrence of the disease till today.

CASE PRESENTATION

In 2013, a 65-year-old man presented to us with complaints of a slow increase in size of his abdomen of 2 years' duration which had initially started off as an eversion of his umbilicus. Besides some intermittent nausea, there were no associated symptoms at presentation. The patient had no comorbidities and no significant medical or surgical history. Clinically, it appeared as if his abdomen was gravid! On palpation, the superior border was thought to be 2-finger breadths below the ribcage and the inferior border was difficult to ascertain.

INVESTIGATIONS

An ultrasound scan was performed initially, which showed a thick-walled septated complex cystic mass arising from the pelvis. This was further investigated by a CT scan of thorax–abdomen and pelvis

which demonstrated a 32×18×25 cm³ cystic mass arising from the left upper quadrant, displacing all intra-abdominal structures (see figure 1); the pancreas was to the level of the iliac crest, and the head of the pancreas was situated anterior to the inferior pole of the kidney along with the splenic artery (see figure 2). No significant lymphadenopathy or bone lesions were seen. The inferior vena cava and the aorta were free from this mass. This mass was likely to represent a benign disease.

Haematological and biochemical investigations revealed only a mild normocytic normochromic anaemia (haemoglobin 10 g/dL). However, a few weeks prior to the surgery, the patient reported 5 episodes of melaena and his Hb was found to be 7.5 g/dL and the patient was transfused with 1 unit of packed red cells. A gastroscopy was carried out which showed no signs of bleeding. The procedure was expectedly challenging due to the extrinsic pressure the mass exerted on the upper gastric anatomy. The mass was not biopsied preoperatively due to the high liquid content of the cyst which could result in accidental rupture.

Postoperatively, the cytology of the intratumorous fluid revealed only leucocytes and red cells. Fluid culture showed no growth of micro-organisms. Histological analysis showed a massive cystic GIST composed by mainly spindle cell with

Figure 1 CT scan of thorax–abdomen and pelvis which demonstrated a 32×18×25 cm³ cystic mass arising from the left upper quadrant.

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Figure 2 CT scan showing the head of the pancreas was situated anterior to the inferior pole of the kidney along with the splenic artery.

a focally storiform and palisading architecture and a mitotic rate of 5 per 50 hpf.

DIFFERENTIAL DIAGNOSIS

The initial work up of this slow-growing soft tissue tumour included sarcoma, desmoid, congenital or germ-line tumours. These were soon dismissed after the CT scan, as it appeared that this mass arose from the left upper quadrant.

The new working differentials included a pancreatic neuroendocrine tumour. This was unlikely with normal amylase and lipase levels and negative carcinoembryonic antigen (CEA) and CA19.9 tumour markers.

This left us with one working diagnosis of an exophytic gastric growth which could be a GIST.

TREATMENT

After an elaborate comprehensive discussion at the regional multidisciplinary meeting (MDT) involving both, upper and lower GI surgeons, it was best thought to surgically excise this cystic mass of uncertain origin, at the earliest.

The decision of the MDT was communicated to the patient and a clinic appointment was made to discuss the nature of surgery at length. The risk of massive bleeding was explained to the patient as it was evident on the CT scan that vessels such as the splenic artery were posterior to the mass, which would

mean if there was any accidental injury to these vessels, gaining haemostatic control would pose a challenging feat. Additionally, the patient was consented for major resections such as gastric, splenic, pancreatic and bowel, should the need arise.

A three-session theatre list along with a cell saver was organised for this case, with 4 units of blood.

The surgery involved a full midline incision. A giant mixed composition GIST arising from the posterior wall of the stomach extending to the pelvis was seen. The upper GI vasculature was found to be dilated and congested with evidence of collaterals. Careful dissection of the large and small bowel off the mass was made. The cystic mass was decompressed by drainage of the fluid and then excised off the pancreas and stomach. A total of 7L of brown fluid was aspirated and 3 kg of solid mass of the cystic lesion was excised.

The pancreas was found to be horse-shoe shaped around the bifurcation of the aorta in the pelvis. The pancreas was fixed to the left upper quadrant via sutures. The splenic vessels were not salvageable from the mass, hence a splenectomy was performed. Finally, a sleeve gastrectomy was performed and an extensive wash out of the abdomen was done. Two-wide bore intra-abdominal drains were left in situ. An estimated total of 5 L blood was lost, 2 L of which were recovered by the cell saver.

The cystic mass along with the fluid was sent for cytology, histopathology studies, microbiology, biochemistry and immunochemistry.

The patient was shifted to the intensive treatment unit due to the nature of the surgery and possible cardiovascular compromise after excision of an impressive cyst which exerted great cardiovascular strain to the abdominal vessels.

The patient was discharged 9 days after surgery. The nutritional team were closely involved in the management of this patient due to the 12% loss of weight after accounting the excision of the cyst and sleeve gastrectomy.

OUTCOME AND FOLLOW-UP

Immunochemistry studies and histopathology studies confirmed the intraoperative suspicion of a GIST. This cystic GIST was diffusely positive for CD117 and DOG1 and very focally positive for SMA (smooth muscle antigen) and desmin. S100, NFP and MNF116 were negative. Histology confirmed a normal spleen and moderate to severe chronic inflammation of the gastric mucosa with no residual tumour. Due to the characteristics of the GIST and moderate to high recurrence risk, it was thought to be best to start the patient on targeted therapy using imatinib for 3 years.

Interestingly, a follow-up CT scan was reported with findings of what seemed to reflect a nodular lesion in the left upper quadrant likely to represent a residual/recurrent disease. On reviewing the images by the operating surgeon, this was soon clarified at the MDT as the suture repair to fix the pelvic pancreatic tail back up to the abdomen or 'pancreatopexy'—where by the floppy unanchored pancreas was fixed into its near normal anatomical site. This procedure has not been described elsewhere (see [figure 3](#)).

The chemotherapy course concluded in September 2016. As of May 2017, the patient is still disease-free and enjoys a normal lifestyle. He is still under a regular follow-up. He has developed a small incisional hernia from his laparotomy, which he prefers not getting operated as he is symptom-free.

DISCUSSION

GISTs are rare soft tissue cancers accounting for only 1.5/100 000 cancers per year in Europe.^{1,2} Patients are normally diagnosed around at the age of 60–65 years. The exact aetiology of GIST remains unknown; however, it has been found that patients may

Figure 3 The 'pancreatopexy' creating a nodular-like lesion on imaging.

have another neoplasm suggesting a strong link with inherited increased susceptibility of malignancy.^{3 4} GISTs originate from the intestinal pacemaker cells of Cajal, often with a KIT tyrosine kinase mutation. Patients may be asymptomatic or may have a spectrum of symptoms such as abdominal mass/pain, bleeding and anaemia, or non-specific symptoms such as night sweats and fever. GISTs tend to be diagnosed late when patient presents with vague symptoms. Incidental GISTs are not uncommon and usually require surveillance and no other treatment. Surgical treatment is the most definitive treatment for symptomatic GISTs and 60% of the patients are cured after complete excision. A risk of disease recurrence exists in 50% of patients and targeted therapy using imatinib is often used as an adjuvant therapy in patients with moderate-risk to high-risk profiles based on tumour size and high mitotic activity.^{5–7} Cystic GISTs are rare lesions that often present a diagnostic dilemma. Moreover, cystic GISTs with malignant potential tend to undergo central cystic degeneration, necrosis and liquefaction. The metastatic lesions of this would also present itself as cystic lesions, which may be mistaken as a primary benign cystic lesion of the organ where

the metastasis has occurred.⁸ Several case reports have been published in peer-reviewed journals describing similar diagnostic conundrums.^{7 9–11} It is strongly encouraged to include GIST as an early differential diagnosis when the exact aetiology of an abdominal mass or a symptomatic abdomen is unknown.

Learning points

- ▶ Gastrointestinal tumours may present in a variety of symptoms and it is important to have an early suspicion of gastrointestinal stromal tumour (GIST) to aid in a timely diagnosis of the condition.
- ▶ Any cystic or solid mass arising from the left upper quadrant should be evaluated for a GIST.
- ▶ Early adjuvant therapy in high-risk patients prevents recurrence of the disease.
- ▶ A multidisciplinary team approach and a multimodal therapy should be emphasised in every case.

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