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Case Report

Perforated small bowel GIST in an immunocompromised male

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ARTICLE INFO

Article history:

Received 4 December 2018

Revised 27 January 2019

Accepted 28 January 2019

Available online 8 February 2019

Keywords:

GIST

Immunocompromised

HIV

Bowel perforation

ABSTRACT

Gastrointestinal (GI) stromal tumor is a relatively rare tumor of the GI tract, with estimated prevalence of 2%, which arises from the interstitial cells of Cajal. Common presentations range from asymptomatic to abdominal pain, bloating, GI bleeding, and anemia. Less-common signs include obstruction and peritonitis. In this case, a young immunocompromised patient experienced a changing symptomatic spectrum with an enlarging abdominal mass, which ultimately perforated, leading to acute peritonitis.

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Case report

A 19-year-old male with past medical history significant for HIV and noncompliance with antiretroviral therapy presented to the emergency department (ED) for lower abdominal pain and nausea. Computed tomography (CT) at that time identified a 4-cm mass of the ileum (Fig. 1). Less than 3 years later, the patient returned to the ED for GI bleeding. A CT showed that the mass had enlarged to 15 cm with internal heterogeneous composition and significant mass effect (Figs. 2 and 3). Before CT-guided biopsy the mass spontaneously developed internal gas (Fig. 4). Biopsy sample showed necrotic debris and enteric bacteria.

Several days later, patient came back to the ED for severe abdominal pain, nausea, and vomiting. CT at this time demonstrated marked central necrosis and air-fluid levels in the mass, but no free intraperitoneal air (Figs. 5 and 6). Over

the course of the next 12 hours, the patient developed peritoneal signs prompting an emergent exploratory laparotomy. The mass was found to have perforated and a fistulous connection to the distal small bowel lumen was noted. The tumor and affected segment of ileum were resected. Pathology identified the mass to be a gastrointestinal stromal tumor (GIST).

Discussion

Gastrointestinal stromal tumors, or GISTs, are the most common mesenchymal tumors of gastrointestinal tract, representing approximately 2% of gastrointestinal tumors [1,2]. While the vast majority of patients remain asymptomatic (70%) with tumors discovered incidentally, those with symptoms typically present with symptoms related to tumor size and location, including early satiety, bloating, and

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<https://doi.org/10.1016/j.radcr.2019.01.014>

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Fig. 1 – Axial oral and IV contrast-enhanced CT abdomen and pelvis of the patient at initial presentation, age 19. A 4 x 3 x 2 cm pelvic mass (arrow) is abutting the ileum and rectum.



Fig. 3 – Coronal CT abdomen and pelvis shows the heterogeneous, 15 x 12 x 11 cm mass (arrow).



Fig. 2 – Axial IV contrast-enhanced CT abdomen and pelvis of the patient at second presentation, age 21. The mass is now 15 x 12 x 11 cm (arrow) and is heterogeneous with significant mass effect.



Fig. 4 – Axial contrast-enhanced CT of the patient prior to percutaneous biopsy demonstrated that within a few days, the mass spontaneously developed gas and air-fluid levels (arrow).

GI bleeding [3,4]. There is no gender predilection with GIST's tending to occur in late middle-age individuals, with 75% occurring in patients >50 years of age [2]. While any region of the gastrointestinal tract may be involved, they are most often seen in the stomach (40%-70%), followed by the small intestine (20%-25%) [3]. Extraintestinal GISTs are rare, but may be seen in the gallbladder, liver, pancreas, peritoneum, adrenal glands, and distantly in the pelvis [3]. While there have been several cases of malignant GIST in pediatric and adult patients with HIV/AIDS, no clear associations have yet been found [5].

CT remains the diagnostic imaging study of choice, is sensitive for detecting lesions >2 cm in size, and aids in the staging and treatment response [3,4]. Additional imaging choices include magnetic resonance imaging and ultrasound, both of which aid in the detection of liver metastases [6,7]. Magnetic resonance imaging is helpful in the delineation of rectal GIST

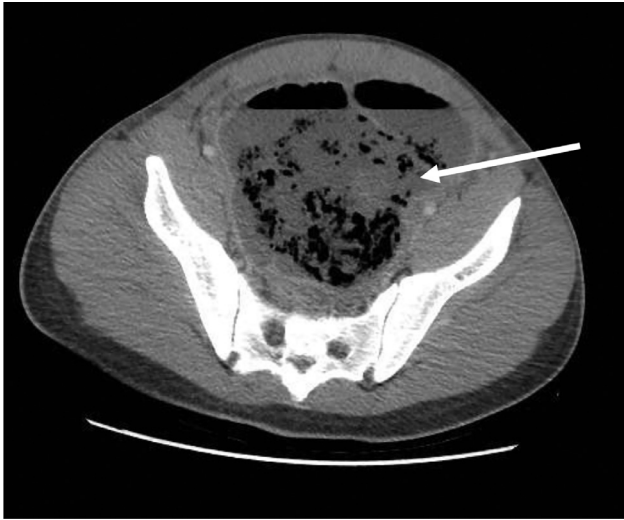


Fig. 5 – Axial contrast-enhanced CT abdomen and pelvis of the patient at third presentation, 3 days after biopsy, shows the heterogeneous mass with internal gas and fluid (arrow). No free intraperitoneal air is present. Scan was obtained prior to exploratory laparotomy.

and identifying regions of hemorrhage/necrosis, while endoscopic ultrasound may be particularly helpful in identifying subtle submucosal GISTs [6,8]. Fluorodeoxyglucose positron emission tomography (FDG-PET) remains useful in the detection of occult GIST as well as monitoring treatment response, although cost and accessibility make it less practical for routine use [9].

Important imaging findings include the primary mass, which is typically >5 cm at presentation, which is often a hypervascular and heterogeneously enhancing exophytic lesion with intratumoral vessels on contrast-enhanced imaging [6]. Most tumors are well-circumscribed with smooth or lobulated margins, with irregularity of the margins suggestive of malignancy [6]. GISTs may locally invade, and if present, metastatic lesions are most often to liver or peritoneum in advanced disease [6]. Fistulization and ulceration may be seen, potentially with air-fluid levels or oral contrast extravasation [6]. Despite the typical large size of the tumor at presentation, GISTs rarely lead to bowel obstruction, although they may displace adjacent structures [6].

Treatment is generally complete surgical resection, the tyrosine kinase inhibitor, Imatinib, is effective in over 70% of cases with unresectable or advanced disease [6]. GISTs often recur, with median recurrence of 2 years, and on surveillance imaging, development of enhancing nodule within a treated area is most suggestive of recurrence [6].

The case of GIST tumor in this patient is unique as it demonstrates the temporal evolution of the mass, from a small asymptomatic mass to a large necrotic mass with fistulous formation and serosal perforation. Additionally, this 19-year-old patient does not fall in the typical demographic of a patient with GIST. Whether this is related to the patient's immunocompromised state is unclear, at present, there is no known connection between HIV-positive status and GIST, although it has been documented in a prior case report.

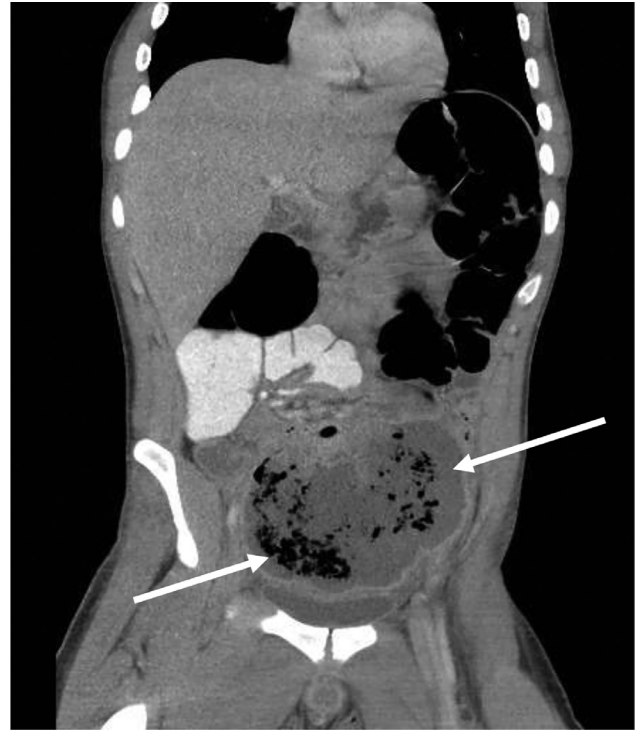


Fig. 6 – Coronal CT shows the heterogeneous mass with internal gas and fluid (arrows). No free intraperitoneal air is present.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.01.014](https://doi.org/10.1016/j.radcr.2019.01.014).

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