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Surgery

NON-APPENDICULAR RIGHT ILIAC FOSSA PAIN, CASE OF RUPTURED GIST: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT Gastrointestinal Stromal Tumors (GISTs) are rare neoplasms originating from the interstitial cells of Cajal, primarily affecting the gastrointestinal tract. This case report details a 35-year-old male presenting with right iliac fossa pain, initially suspected to be appendicitis. Upon surgical exploration, a ruptured GIST was discovered in the proximal ileum. The patient underwent limited resection and anastomosis, with histopathology confirming a malignant GIST. This report draws attention to the paucity of research on ruptured Gastrointestinal Stromal Tumors (GISTs), highlighting the tendency to neglect or underestimate the risks associated with these tumors. The study emphasizes the intricacies involved in managing ruptured GISTs, stressing the importance of surgical intervention and targeted therapeutic strategies.

KEYWORDS : Gastrointestinal Stromal Tumors (GISTs); Interstitial cells of Cajal (ICC); Torricelli-Bernoulli sign; Locoregional therapy; Metastases; Tyrosine kinase inhibitors

INTRODUCTION:

The right iliac fossa, a defined anatomical region situated in the lower right quadrant of the abdomen, is a location where a diverse range of pathological conditions can manifest, often leading to acute or chronic discomfort for the patient. Accurate diagnosis of the underlying cause of right iliac fossa pain is crucial, as it can guide appropriate treatment and prevent potential complications. [1]

One of the most common causes of right iliac fossa pain is acute appendicitis, a condition characterized by inflammation of the appendix. In such cases, the pain may initially be diffuse but eventually localize to the right lower quadrant, often accompanied by other symptoms such as nausea, vomiting, and fever.[2] Atypical presentations, where the appendix is located in an anomalous position, can make diagnosis more challenging, emphasizing the importance of imaging modalities like computed tomography to identify the cecum and locate the appendix. [3]

Some of the alternative diagnoses include Meckel's diverticulitis, Mesenteric adenitis, Infective colitis (typhilitis), Terminal ileitis, Right-sided diverticulitis, Intussusception, Renal colic/stone, Cholecystitis and ectopic pregnancy. [4] Although rare, Gastrointestinal Stromal Tumors (GISTs) have been reported to present with right-iliac fossa pain or mass.[5]

GISTs arise from the interstitial cells of Cajal (ICC), which are specialized cells located in the myenteric plexus, a network of nerve fibers that controls gut motility; and can range from small, benign tumors to large, malignant ones. They are typically located within the GI tract, most commonly in the stomach (50-60%) and small intestine (20-30%). However, they can also arise outside the GI tract in mesentery, retroperitoneum and omentum.[6] Although a majority of these arise from non-hereditary mutations in KIT or PDGFRA genes leading to uncontrolled tumour cell proliferation, a significant proportion of cases 10-15% of adult GISTs and 85% of pediatric GISTs lack these mutations.[7]

Adults aged 40 and above are the most affected, with an equal distribution between men and women.[8] When it comes to ethnic backgrounds, Blacks have the highest incidence rate.[7]

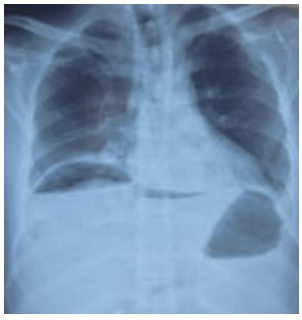
Most commonly they present with early satiety and abdominal pain, but in rare occasions may also present with complications such as bleeding or signs of intestinal obstruction.[7]

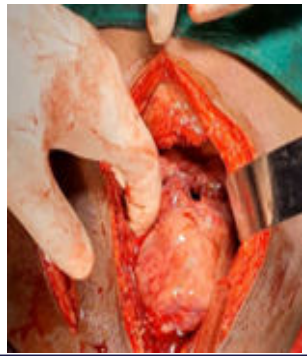
GISTs with high mitotic rates (exceeding 5 per 10 high-power fields) or large size (over 10 cm) are classified as histologically malignant due to increased risk of recurrence and metastasis.[8]

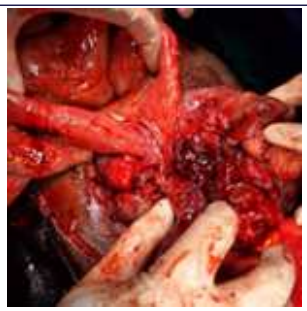
While endoscopic and percutaneous biopsy confirms the diagnosis, additional diagnostic tools may include imaging studies like CT, MRI, PET; Immunohistochemistry (IHC) testing for KIT, DOG1, or other markers and Molecular analysis for genetic mutations. The precise diagnosis of these tumours has gained significant importance with the advent of Imatinib mesylate (Gleevec/Glivec), a targeted tyrosine kinase inhibitor that selectively targets KIT-positive tumors, revolutionizing the treatment of unresectable and metastatic GISTs[9]

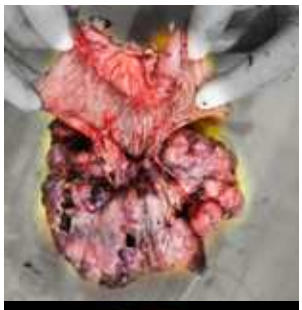
Optimal treatment outcomes for localized GISTs are achieved through complete surgical excision with clear margins and minimizing tumor rupture or spillage during resection. Whereas for patients with locally advanced GISTs, metastatic disease and aggressive tumor behavior, a combination of chemotherapy and surgery may offer improved outcomes. For GIST cases that are refractory or poorly responsive to traditional tyrosine kinase inhibitor therapy, immunotherapy has emerged as a novel and promising therapeutic approach under active investigation.[7]

CASE REPORT









A 35-year-old male manual laborer by occupation, a migrant from Bihar presented to the casualty with complaints of pain abdomen since 2 days – initially in Right iliac fossa, now with generalized, dull aching type which aggravates on exertion and relieves on medication and lying still, associated with multiple episodes of vomiting, bilious in nature and not blood tinged and fever with chills since 1 day, no h/o any obstipation and no previous h/o similar episodes in the past or any past history of surgeries in the past. Had no known comorbidities and not on any analgesic abuse or steroid use.

On examination, patient was concious and oriented, with tachycardia and hypotension and abdomen is distended and no visible lump/peristalsis/scars/sinuses/visible veins over the abdomen on inspection and diffuse abdominal tenderness with guarding and board like rigidity. Bowel sounds were absent. Hernial orifice was normal. Per Rectal examination had collapsed rectum and gloved finger stained with stools.

Erect Xray – revealed air under diaphragm and USG was requested suspecting appendicular perforation, USG showed – mild ascites with collapsed bowel loop in right and left iliac fossa, with appendix not visualized.

With a strong suspicion of Peritonitis secondary to perforated appendicitis, right Paramedian Laparotomy was undertaken under Sub-arachnoid block. On opening the peritoneum, brownish aspirate ? Hemorrhagic fluid could be noted, and further examination showed a large mass in the RIF. Hence, proceeded with midline laparotomy incision with conversion to GA. A large well circumscribed nodular mass could be noted, arising from the wall of proximal ileum with area of necrosis. Limited resection and anastomosis was undertaken and post op specimen when cut open showed no luminal breech.

Patient was shifted out in stable condition to post op ward for monitoring. POD-1 was uneventful and the patient had hypokalemia (K=3.4) drain output was 10 ml serosanguinous and RT aspirate of about 300 ml bilious. Patient was mobilized on POD-2 and wound inspected showed to be healthy. Patient had passed flatus on POD-2, POD-3 Ryles tube was removed and patient was started on sips of water. As the drain had no significant output the drain was removed on POD-5. Patient was advised to take high protein diet. Patient absconded from the ward on POD-7 and could not be traced. Patient was planned to be sent to higher centre after HPE report.

HPE: Malignant tumor arising from muscularis propria, tumor arranged in sheets and interlacing fascicles, with tumor cells being spindle shaped and abnormal mitotic figures of 5-6/hpf — Malignant GIST or Leiomyosarcoma with IHC confirming it to be GIST.

DISCUSSION:

Gastrointestinal stromal tumors are a type of rare and complex neoplasm that originate from the interstitial cells of Cajal, which are responsible for regulating the motility of the gastrointestinal tract. These tumors can occur throughout the digestive system, with the stomach being the most common site of occurrence.[8] Although GISTs are generally considered to be indolent tumors, a subset of these can exhibit aggressive behavior, leading to rupture and subsequent poor prognosis. Notably, a distinct subtype of GISTs is the succinate dehydrogenase-deficient variety, which has been observed to occur in association with certain rare syndromes, such as the Carney triad and the Carney-Stratakis syndrome.

Tumor rupture is a significant risk factor for predicting recurrence after macroscopically complete resection of gastrointestinal stromal tumors (GISTs) and is a basis for recommending defined intervals or even lifelong adjuvant therapy with imatinib, as outlined in guidelines. However, there is no universally agreed-upon definition of "tumor rupture," leading to considerable variability in its reported incidence across different studies.[11]

Torricelli-Bernoulli sign occurs when internal necrosis of a submucosal tumor (leiomyosarcoma or GIST) is associated with ulceration into the intestinal lumen, releasing a stream of air bubbles that can be visualized by diagnostic imaging [10] or infection of the peritoneal contamination by gas producing organism might be the cause of air under the diaphragm in this case as post op specimen cut section and the histopathology showed no luminal breech for air to have escaped to cause air under the diaphragm.

Surgery remains the cornerstone of treatment for primary, localized GISTs, with the goal of complete R0 resection. However, in the case of a ruptured GIST, the risk of tumor seeding and peritoneal dissemination increases significantly, leading to a higher likelihood of incomplete resection and disease recurrence. Systemic therapy with tyrosine kinase inhibitors, such as imatinib and sunitinib, has revolutionized the management of advanced and metastatic GISTs, but their role in the setting of a ruptured tumor is less well-established.[12] Preoperative treatment with imatinib may play a role in cytoreducing the tumor burden and facilitating a more complete surgical resection in patients with large or locally advanced GISTs.[13] Additionally, the use of locoregional therapies, such as radiofrequency ablation or transarterial embolization, has shown promise in the management of GIST metastases, particularly in cases where surgical resection is not feasible.[14]

CONCLUSION:

Gastrointestinal stromal tumors (GISTs) are relatively rare solid tumors, making up less than 5% of all tumors in the digestive tract. They are often discovered incidentally during imaging conducted for other health concerns or occasionally presents with complications. Typically, these tumors are only a few centimeters in size. Larger GISTs can exert pressure on surrounding structures, causing symptoms such as early satiety, changes in bowel habits, difficulty swallowing (dysphagia), jaundice, or bowel obstruction. The epidemiology of ruptured GISTs remains a topic of ongoing investigation, with limited data available in the literature. Surgery is the primary treatment for localized, primary GISTs, aiming for a complete R0 resection. However, when a GIST ruptures, the risk of tumor seeding and spread within the peritoneum rises significantly, which increases the chances of an incomplete resection and a higher likelihood of disease recurrence.

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