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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/16190
DOI URL: http://dx.doi.org/10.21474/IJAR01/16190

RESEARCH ARTICLE

A RIGHT ILIAC FOSSA MASS - A PERFORATED ILEAL GASTRO-INTESTINAL STROMAL-TUMOUR

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Manuscript Info

Manuscript History
Received: 29 November 2022
Final Accepted: 30 December 2022
Published: January 2023

Abstract

GASTRO-INTESTINAL STROMAL-TUMOURs (GASTRO-INTESTINAL STROMAL-TUMOURs) are tumours of the gut found mostly in stomach and small intestine. The complications are Gastrointestinal (GI) bleeding, obstruction, pain and rarely perforation. We are reporting an abnormal presentation of GASTRO-INTESTINAL STROMAL-TUMOURS presenting as an acute abdomen pain with Right Iliac Fossa (RIF) mass in 57-year-old male. Contrast Enhanced Computed Tomography (CECT) of abdomen revealed a peripherally enhancing encapsulated abscess in RIF close to the base of caecum and surrounding peritonitis suggesting a caecal perforation. On laparotomy, a perforated ileal GASTRO-INTESTINAL STROMAL-TUMOURS was identified. Pathological examination confirmed the tumour to be a GASTRO-INTESTINAL STROMAL-TUMOUR of spindle cell type, further reiterated by immunohistochemistry. Our case report emphasizes, GASTRO-INTESTINAL STROMAL-TUMOUR are a rare differential diagnosis of RIF mass, and to have high clinical suspicion when a similar case is encountered in an emergency room, keeping in mind the poor outcome caused by delay in appropriate management of the disease.

Introduction:-

Case Report:

A 57-year-old male presented to the Emergency room with c/o diffuse lower abdominal pain for 3 days with few episodes of haematochezia in the past 1 day. On examination, she had pallor and tachycardia. On examination of abd0men, there was tenderness, mild guarding, rebound tenderness and an ill-defined mass in the right iliac fossa (RIF). On per rectal examination, finger blood stained. The laboratory investigations revealed haemoglobin of 9 gm% and Total WBC count of 13826 cells/mm3. CXR and abdomen X-rays were normal. Ultrasound abdomen revealed ill-defined mass in RIF with minimal free fluid and probe tenderness. CONTRAST ENHANCED COMPUTED TOMOGRAPHY of abdomen showed a peripherally enhancing, encapsulated abscess in RIF, close to the base of caecum and surrounding peritonitis, suggesting a sealed caecal perforation. [Fig – 01]

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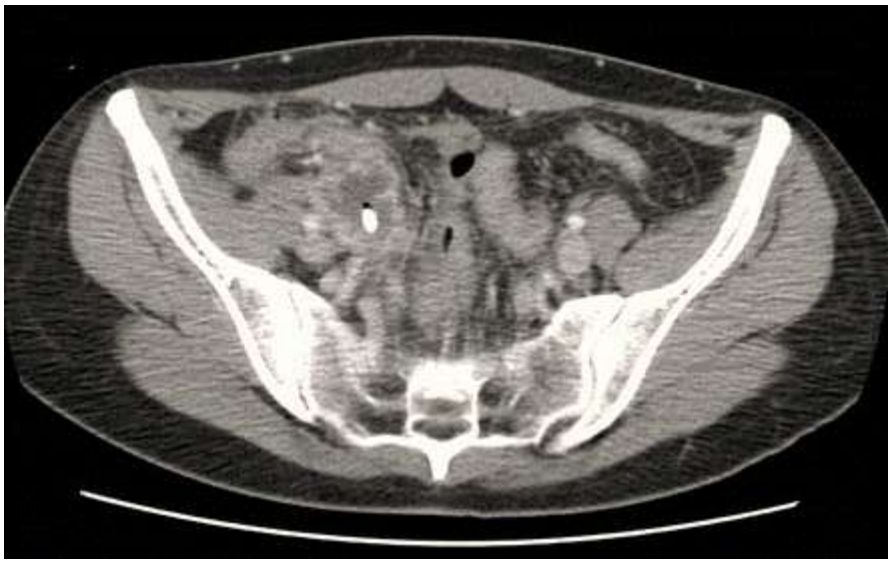
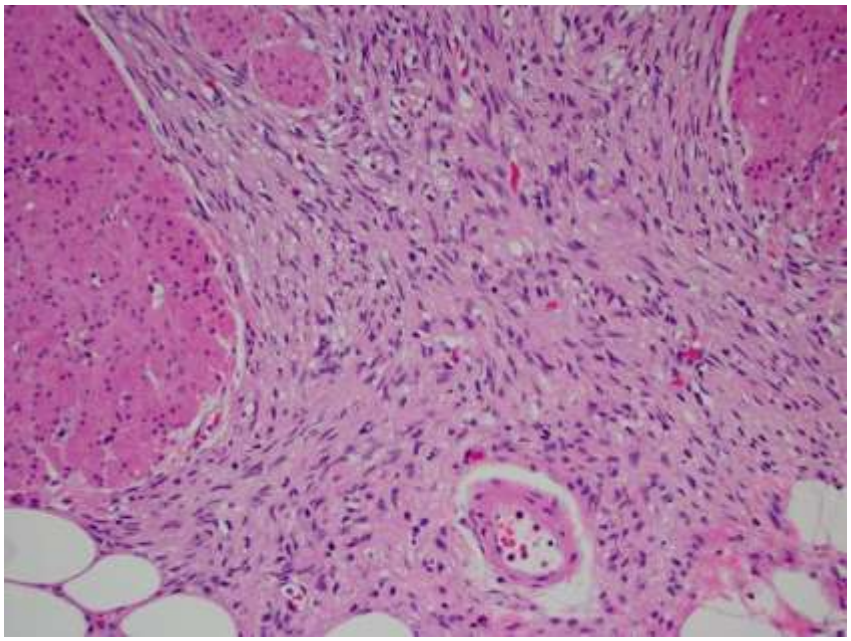


Fig 1 :- CECT abdomen showing an encapsulated abscess in RIF, close to the base of caecum and surrounding peritonitis, suggesting a sealed caecal perforation.

Intra – Operative findings:

He was taken up for an emergency laparotomy. It showed that a part of the omentum and the small bowel were adherent to the caecum and nearby abdominal wall. They were slowly dissected by blunt dissection. This revealed a perforated small bowel tumour of maximum diameter 7 cm – 8 cm at the terminal part of the ileum nearly, 3 cm proximal to the ileocecal junction [Fig – 2]. Rest of the abdomen was normal. He underwent Right Hemicolectomy. The abdomen was washed with 2L of normal saline and incision was closed in layers.

Pathological findings:
Histo-Pathological Examination revealed a 7 cm x 4 cm x 4 cm submuc0sal mass extending into the antimesenteric border of ileum. Microscopic examination showed a low grade, intermediate risk, pT3 pNx GASTRO-INTESTINAL STROMAL-TUMOUR. CD117 being positive confirmed the diagnosis of GASTRO-INTESTINAL STROMAL-TUMOUR



Postoperative course:
His postoperative period was uneventful. He was discharged on POD 10 after suture removal. He was started on adjuvant imatinib therapy to prevent tumour recurrence.

Discussion:-

GASTRO-INTESTINAL STROMAL-TUMOUR are tumours of mesenchymal origin that arise from interstitial cells of Cajal in GIT. They constitute 0.1%-3% of all Gastrointestinal Tract (GIT) tumors [1]. GASTRO-INTESTINAL STROMAL-TUMOURs typically occur in the elderly around the sixth decade of life [2]. They can occur anywhere in the GIT but mostly observed in the stomach (50%) and small intestine (25%). GASTRO-INTESTINAL STROMAL-TUMOURs generally have an indolent course and are diagnosed incidentally. Small sized tumours remain silent while large sized tumours present as large abdominal masses with clinical features unrelated to the disease, so most metastasize at the time of presentation [2,3]. The complications are GI bleeding (40%), intestinal obstruction, abdominal pain and very rarely perforation [2].

GASTRO-INTESTINAL STROMAL-TUMOUR as a content of Meckel's diverticulum has been reported in seven cases [4] and as abdominal cocoon in a single case [5]. Most common complication of GASTRO-INTESTINAL STROMAL-TUMOUR is bleeding [2] but they have also presented rarely as perforation which has been documented in seven cases [6]. There are 2 documented cases till now of GASTRO-INTESTINAL STROMAL-TUMOUR presenting as RIF mass, one a jejunal GASTRO-INTESTINAL STROMAL-TUMOUR masquerading as an appendicular mass [7] and another a cecal GASTRO-INTESTINAL STROMAL-TUMOUR presenting as a diffuse swelling in RIF and right lumbar regions [8]. In our case, a perforated terminal ileal GASTRO-INTESTINAL STROMAL-TUMOUR presented as a RIF mass with lower GI bleed. This combination of presentation is yet to be reported.

The pathological diagnosis is based on the histological examination of the tumour and IHC staining is required to confirm the diagnosis [9]. The following table shows the IHC staining pattern of GASTRO-INTESTINAL STROMAL-TUMOUR [Table/Fig-5] [2,5,10-13].

Table/Fig-5:- Positivity rates for various immunohistochemical markers in GASTRO-INTESTINAL STROMAL-TUMOUR.

Marker	Positive rate (%)
CD 117	94-98
CD 34	60-80
SMA	20-30
S100	10
Desmin	<5

Recently identified, antibody against DOG1 (discovered on GASTRO-INTESTINAL STROMAL-TUMOUR) is reported to be positive for 85%-95% of CD117-positive GASTRO-INTESTINAL STROMAL-TUMOURs and in 30%-36% of CD117–negative GASTRO-INTESTINAL STROMAL-TUMOURs. However, it can't differentiate between KIT/PDGFRA mutant and wild-type GASTRO-INTESTINAL STROMAL-TUMOUR. Hence, DOG1 immuno-staining may be helpful to identify tumours which cannot be diagnosed based on C_Kit immunohistochemistry [11].

Fletcher CD et al., proposed a classification system to prognosticate GASTRO-INTESTINAL STROMAL-TUMOUR and has been widely accepted and followed today [Table/Fig-6] [14]. The mitotic count is the most vital prognostic factor [10,12].

Table/Fig-6:- Classification system by Fletcher et al.

Risk of malignancy	Size of tumour (cm)	Mitotic counts (/50HPF)
Very low	<2	<5 / 50
Low	2–5	<5
Intermediate	<5	6– 10
	5 – 10	<5
High	>5	>5
	> 10	Any counts
	Any size	>10

The preferred treatment of choice for GASTRO-INTESTINAL STROMAL-TUMOURs is R0 surgical excision. A lymphadenectomy is usually not performed, as metastasis to lymph nodes is extremely rare. Patients with perforation have a five-year survival rate of only 24%, in contrast to patients with localized or locally advanced tumours where the five-year survival rate is 46%. The reason for this difference may be attributed to peritoneal dissemination [13]. Imatinib, a selective tyrosine kinase inhibitor therapy which has significantly improved the overall survival in patients with advanced disease. Adjuvant therapy should be considered for four years, in patients undergoing R0 resection for primary disease and perforation as well [15].

Conclusion:-

We presented this case of an acute abdomen with a RIF mass that was incidentally diagnosed to be a perforated GASTRO-INTESTINAL STROMAL-TUMOUR arising from the ileum. Apart from the usual causes of RIF mass, a perforated GASTRO-INTESTINAL STROMAL-TUMOUR of this kind should also be considered when older patients present with such clinical features. A high degree of clinical suspicion and prompt management is required in view of the high morbidity rates from delayed diagnosis of this disease.

Financial or Other Competing Interests
None.

References:-

[1] Connolly EM, Gaffney E, Reynolds JV. GASTRO-INTESTINAL STROMAL-TUMOURs. Br J Surg. 2003;90(10):1178–86.
[2] Miyata S, Bliss DW. A gastrointestinal stromal tumor found in perforated Meckel's diverticulum. Surg case reports. Springer. 2016;2(1):67.
[3] DeMatteo RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF. Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. Ann Surg. 2000;231(1):51–58.

[4] López-TomassettiFernández EM, Hernández Hernández JR, Nuñez Jorge V, Usha M. Perforated gastrointestinal stromal tumor in Meckel's diverticulum treated laparoscopically. Asian J Endosc Surg. 2013;6(2):126–129.
[5] Kumar V, Rau RA, Kamath S. Perforated GASTRO-INTESTINAL STROMAL-TUMOUR in Jejunum - A Rare Cause of Abdominal Cocoon. J ClinDiagn Res. 2014;8:132–133.
[6] Skipworth J, Fanshawe A, West M, Al-Bahrani A. Perforation as a rare presentation of gastric GASTRO-INTESTINAL STROMAL-TUMOURs: a case report and review of the literature. Ann R CollSurg Engl. 2014;96(1):01–05.
[7] Nancharaiah P, Venkateswarlu MC, Aishwarya M. GASTRO-INTESTINAL STROMAL-TUMOUR in Rif masquerading as appendicular mass. IOSR J Dent Med Sci. 2016;15:2279–2861.
[8] Sreevathsa MR. Caecal gastrointestinal stromal tumor with perforation and obstruction. Indian J SurgOncol. Springer. 2012;3(4):311–313.
[9] Grover S, Ashley SW, Raut CP. Small intestine gastrointestinal stromal tumors. CurrOpinGastroenterol. 2012;28:113–23.
[10] Judson I. GASTRO-INTESTINAL STROMAL-TUMOURs (GASTRO-INTESTINAL STROMAL-TUMOUR): biology and treatment. Ann Oncol. 2002;13(Suppl 4):287–89.
[11] Bucher P, Taylor S, Villiger P, Morel P, Brundler MA. Are there any prognostic factors for small intestinal stromal tumors? Am J Surg. 2004;187:761–66.
[12] Liegl B, Hornick JL, Corless CL, Fletcher CD. Monoclonal antibody DOG1.1 shows higher sensitivity than kit in the diagnosis of gastrointestinal stromal tumors, including unusual subtypes. Am J SurgPathol. 2009;33:437–46.
[13] Crosby JA, Catton CN, Davis A, Couture J, O'Sullivan B, Kandel R, et al. Malignant gastrointestinal stromal tumors of the small intestine: a review of 50 cases from a prospective database. Ann SurgOncol. 2001;8(1):50–59.
[14] Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: A consensus approach. Hum Pathol. 2002;33(5):459–465.
[15] Koo DH, Ryu MH, Kim KM, Yang HK, Sawaki A, Hirota S, et al. Asian Consensus Guidelines for the Diagnosis and Management of Gastrointestinal Stromal Tumor. Cancer Res Treat. 2016;48(4):1155–66.