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***Case Report***

# ROUND CELL NEOPLASM (GIST) OF TERMINAL ILEUM - A CASE REPORT

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## ABSTRACT

A 50 year old man of North Indian origin presented with an acute tender lump in the right iliac fossa. The lump was initially thought to be an appendicular lump. On exploratory laparotomy, a hard suspicious mass was found in the terminal ileum with mesenteric lymphadenopathy. Frozen section of the lymph node showed reactive hyperplasia and biopsy of ileocaecal respected specimen revealed round cell neoplasm in terminal ileum invading all layers.

***Keyword:*** *Round Cell Neoplasm (Gist), Ileum*

## CASES

A 50 year old man presented with a 5 month history of moderate localized pain in his right lower abdomen that was not radiating to any other site and was associated with nausea and vomiting. The patient had mild pyrexia (temperature 99.4°F). On examination of his abdomen, a well-defined mildly tender localized fixed lump 6x6cm in size was found in the right iliac fossa.

The hemogram showed a total leukocyte count of 7000/cu.mm, with 74% polymorphonucleocytes.

Ultrasonography (USG) of his abdomen revealed an oval-shaped abdominal mass (8.9x3.7cm) in the right iliac fossa, suggestive of an appendicular lump.

• CECT abdomen revealed asymmetric circumferential wall thickening of distal ileal loops measuring 10-15mm thickness and involving about 90mm long segment and adjacent mesenteric lymph nodes measuring 8-12mm in diameter likely mitotic. D/D- lymphoma / adenocarcinoma.

### *Operative Treatment and Chemotherapy Given*

Surgical excision of small bowel tumors remains the recommended therapy.

Both segmental resection and enterotomy/polypectomy have been used for lesion removal.

If the pathology cannot be established at the time of resection, full segmental resection with adequate margins is recommended

No standard regimen demonstrates benefit in an adjuvant or metastatic setting for small-bowel adenocarcinoma.

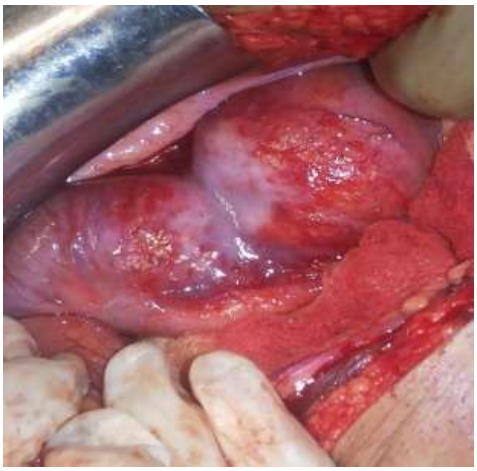
Because of the similarity to CRC, a regimen containing 5-FU with leucovorin may be used. Newer agents as irinotecan and oxaliplatin, may also be used with 5-FU.

### *Biopsy: (Picture: 1 and 2)*

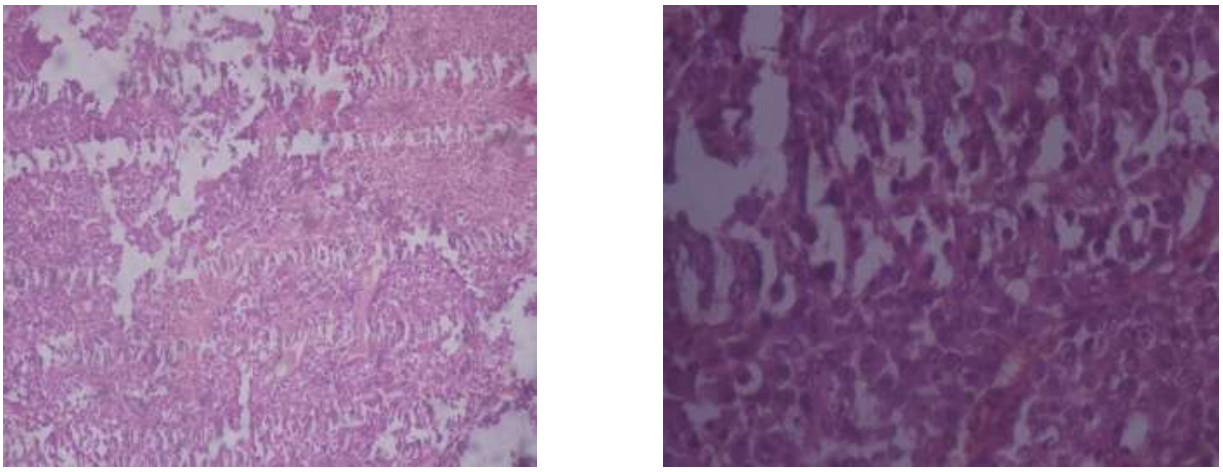
Microscopy of growth in ileum revealed round cell neoplasm invading all layers completely (markers are under study).

Appendix showed lymphoid follicular hyperplasia, Caecum unremarkable and resected end of ileum was free from tumor

Five resected lymph nodes showed reactive hyperplasia.



**Picture 1: Per operative image**



**Picture 2: Histopathological slide images**

## DISCUSSION

GIST occurs marginally frequent in males as compared to females, both in the fifth and sixth decades of life. There is no racial or geographical preponderance. Clinical presentation varies from an incidental radiological finding when a patient is investigated for other symptoms to cases of intestinal obstruction, upper or lower GI bleeding or melena, and also as an emergent idiopathic spontaneous intra-abdominal hemorrhage. As in the case being reported, some may present with a palpable abdominal lump.

Although abdominal ultrasound is often the initial imaging test employed in the investigation of a patient with abdominal pain or mass, the tumor discovered is frequently so large rendering the organ of origin unidentifiable. The sonogram report frequently indicates the presence of a huge mass, often filling the abdomen, of heterogeneous reflectivity and frequent necrosis. CT therefore provides the basis for diagnosis and staging in most patients. Tumors are usually of varying density, and show patchy enhancement after intravenous contrast. Varying degrees of necrosis may be frequently demonstrated within the mass (King, 2005). The CT study will usually provide rapid and reproducible assessment of the size of the primary tumor, as well as its relationship to other structures. Metastatic disease may well be demonstrated at the outset (Burkill *et al.*, 2003). CECT abdomen findings of solid pseudopapillary neoplasm of the pancreas could share the radiological features of GIST (Lakhtakia *et al.*, 2013).

GIST can develop into an enormous size before diagnosis and demonstrate considerable cystic change usually associated with a surrounding rim of the viable enhancing tumor. Necrosis may lead to enteric

fistulation. Calcification within the tumor is occasionally recognized in association with this tumor necrosis.

Small bowel neoplasms are rare. Factors – rapid transit, high turnover rate, alkalinity, high IgA, low bacterial counts Mean age- 60 years Low incidence – India, East Europe. Increased after mid 1980 75% symptomatic neoplasms malignant.MC benign neoplasm – Adenoma/Leiomyoma, MC malignant neoplasm – AdenoCa/Carcinoid tumor, Molecular genetics – kras, 5q, 17q, 18q(DCC), DPC4(SMAD4).

Guidelines indicate that radical surgical resection is the gold standard for localized primary GIST. Increasing cure rates, overall survival and progression-free survival should be the aim of all adjuvant therapy which should be reserved only for patients having significant prognostic indicators for disease recurrence (Casali *et al.,* 2010). The Scandinivian Sarcoma Group study showed that adjuvant imatinib given for three years improved recurrence-free survival compared to a one-year therapy.

Recent studies have shown the risk of recurrence is high if tumor spillage occurs during surgery. The present consensus is that patients who have histological profile of intermediate, moderate, or high risk and those with R1 and R2 (microscopic and macroscopic tumor residue) or tumor rupture should receive long-term adjuvant therapy with imatinib. Nomograms have been developed to predict recurrence-free survival after resection of localized primary GIST tumors. This helps to guide patient selection for adjuvant imatinib therapy

## REFERENCES

**Blackstein ME, Corless CL and Ballman KV *et al.*, (2010).** Risk assessment for tumor recurrence after surgical resection of localized primary gastrointestinal stromal tumor (GIST): North American Intergroup phase III trial ACOSOG Z9001. *Program and Abstracts of the 2010 Gastrointestinal Cancers Symposium; January 22-24, 2010, Orlando, Florida*. Abstract 6.

**Burkill GJ, Badran M, Al-Muderis O, Meirion Thomas J, Judson IR and Fisher C *et al.*, (2003).** Malignant gastrointestinal stromal tumor: distribution, imaging features, and pattern of metastatic spread. *Radiology* **226**(2) 527-532. 10.1148/radiol.2262011880

**Casali PG, Blay JY and ESMO/CONTICANET/EUROBONET Consensus Panel of Experts (2010)**. Gastrointestinal stromal tumours: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Annals of Oncology* **21**(Suppl 5) v98-v102. 10.1093/annonc/mdq208

**Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J and Longley BJ *et al.*, (2002).** Diagnosis of gastrointestinal stromal tumors: A consensus approach. *Human Pathology* **33**(5) 459-465. 10.1053/hupa.2002.123545

**Gokhale U and Pillai GR (2009).** Large Brunner's Gland Hamartoma: A Case Report. *Oman Medical Journal* **24**(1) 41-43.

**Hirota S, Isozaki K, Moriyama Y, Hashimoto K, Nishida T and Ishiguro S *et al.*, (1998).** Gain-of-function mutations of c-kit in human gastrointestinal stromal tumors. *Science* **279**(5350) 577-580. 10.1126/science.279.5350.577

**Hohenberger P, Ronellenfitsch U, Oladeji O, Pink D, Ströbel P and Wardelmann E *et al.*, (2010).** Pattern of recurrence in patients with ruptured primary gastrointestinal stromal tumour. *British Journal of Surgery* **97**(12) 1854-1859. 10.1002/bjs.7222

**Kindblom LG, Remotti HE, Aldenborg F and Meis-Kindblom JM (1998).** Gastrointestinal pacemaker cell tumor (GIPACT): gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. *American Journal of Pathology* **152**(5) 1259-1269.

**King DM (2005).** The radiology of gastrointestinal stromal tumours (GIST). *Cancer Imaging* **5**(1) 150-156. 10.1102/1470-7330.2005.0109

**Lakhtakia R, Al-Wahaibi K, Zahid KF, Malik KA and Burney IA (2013).** Solid pseudopapillary neoplasm of the pancreas: a case report with review of the diagnostic dilemmas and tumor behavior. *Oman Medical Journal* **28**(6) 441-444. 10.5001/omj.2013.122

**Mazur MT and Clark HB (1983).** Gastric Stromal Tumours: Reappriasal of Histogenesis. *The American Journal of Surgical Pathology* **7**(6) 509-519.

**Miettinen M and Lasota J (2001).** Gastrointestinal stromal tumors–definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. *Virchows Archiv* **438**(1) 1-12. 10.1007/s004280000338

**Nakamura N, Yamamoto H, Yao T, Oda Y, Nishiyama K and Imamura M *et al.*, (2005).** Prognostic significance of expressions of cell-cycle regulatory proteins in gastrointestinal stromal tumor and the relevance of the risk grade. *Human Pathology* **36**(7) 828-837. 10.1016/j.humpath.2005.03.012

**Nilsson B, Bümming P, Meis-Kindblom JM, Odén A, Dortok A and Gustavsson B *et al.*, (2005).** Gastrointestinal stromal tumors: the incidence, prevalence, clinical course, and prognostication in the preimatinib mesylate era–a population-based study in western Sweden. *Cancer* **103**(4) 821-829. 10.1002/cncr.20862

**Sakurai S, Oguni S, Hironaka M, Fukayama M, Morinaga S and Saito K (2001).** Mutations in c-kit gene exons 9 and 13 in gastrointestinal stromal tumors among Japanese. *Japanese Journal of Cancer Research* **92**(5) 494-498. 10.1111/j.1349-7006.2001.tb01121.x