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CASE REPORT ON RARE OUTCOME OF A RETROPERITONEAL MASS: MANAGEMENT CHALLENGES AND LESSONS LEARNT

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Background. *Gastrointestinal stromal tumours (GIST) are non-epithelial mesenchymal solid neoplasm with varied presentation. The study reports the case of a retroperitoneal GIST in a 21-year-old male presented with an abdominal lump for six months. The lesion was initially thought to be a retroperitoneal sarcoma. Exploratory laparotomy revealed an abdominopelvic mass covering the entire right side of abdomen and pelvis. The tumour was adherent to the terminal ileum and ascending colon. There were dense adhesions between the retroperitoneum with involvement of the middle third of the right ureter. The tumour was resected with right hemicolectomy with ileotransverse anastomosis. Post-operative histopathology revealed it as high-grade spindle cell type GIST. The patient is presently on post-operative chemotherapy with Imatinib mesylate.*

Objective. *Atypical presentations of GIST are seldom discussed but frequently encountered in clinical practice. This article depicts different challenges the surgeon has to face while diagnosing such atypical entity.*

Methods. *Case report of atypical GIST presenting as retroperitoneal lump.*

Results. *The patient underwent surgical resection and is presently on post-operative chemotherapy with good overall outcome for a one year follow up.*

Conclusions. *GISTs presenting as retroperitoneal lumps are very rare, they should be considered in their differential diagnosis of an atypical retroperitoneal mass.*

KEYWORDS: GIST; atypical presentation; retroperitoneal tumour; abdominal lump.

Introduction

Gastrointestinal stromal tumours (GIST) are intra-abdominal benign mesenchymal neoplasia arising from the interstitial cell of Cajal. The most common sites are the stomach and the intestine. Mostly GIST presents as a slow-growing intra-abdominal mass appreciated as a lump abdomen. They can also present as gastrointestinal bleed or intestinal obstruction. However, there are many incidences where they present atypically. This article is about the diagnostic challenges and analysis of various rare presentations of GIST.

Case Report

A 21-year-old male presented with a 6-month history of lump abdomen in the umbilical and right lumbar region. The lump was insidious in onset and gradually progressive. There was no history of vomiting, constipation or obstipation. He also denies any history of trauma to the abdomen. There were no hepatic or urinary complaints. On clinical examination a nontender, smooth surfaced firm 20×15 cm lump was palpated in the right lumbar region extending to the umbilicus crossing 3 cm lateral to the midline. The lump was fixed to retroperitoneum. There was no palpable hepatosplenomegaly, and digital rectal examination was normal.

The patient underwent extensive preoperative investigation for diagnosis. On admission his haemoglobin was 13.5 g%, Total leukocyte count was 6.200/mm^3 with a normal differential count, platelet count and coagulogram. The random blood sugar was 98 mg%. The renal and liver function tests were normal. The abdomen ultrasonogram revealed large heterogeneous mass of 16×11.4 cm in the pelvis region on the right side. Possibility of Lymphoma / lymphoproliferative disorder should be ruled out. Mild fullness of pelvicalyceal system on right side and mild hepatosplenomegaly presented. Keeping a working diagnosis of a retroperitoneal tumour, a contrast enhanced computerised

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tomography scan revealed 13.2×9.9×13.4 cm round to oval, predominantly isodense lesion in lower abdomen and pelvis (Fig. 1).

On the right side, it appeared to involve the mesentero-peritoneal space. Patchy mesenteric fat stranding was seen. Mass effect with displacement of bowel loops and indentation of urinary bladder was evidenced. Another 3.2×3.1×9.7 cm iso to hyperdense lesion was present predominantly in iliopsoas muscle. An ill-defined isodense lesion of 2.2×1.3×1.2 cm was revealed in subcutaneous tissue of anterior abdominal wall in right paraumbilical region. Multiple sub centimetric sized mesenteric lymph nodes was seen. Posterior end of left lower ribs appeared thickened and showed mild deformity. The tumour was locally aggressive with multiple intramuscular, and subcutaneous enhancing nodular lesion was revealed. Non enhancing hyperdense band like lesions was evidenced in iliacus and paraspinalis muscles. There was an Expansile lesion of the 7th and 8th ribs on the left side. The differential diagnosis of desmoplastic fibroma/fibrous dysplasia of the retroperitoneum was contemplated. Fine needle aspiration cytology from the lesion showed no evidence of malignant cells.

A CT angiogram further revealed the same locally aggressive lesion in right lumbar and iliac region with extension into pelvis. The arterial supply of the lesion was seen to arise from hypertrophic ileo-colic-right colic trunk. The diameter of ileo colic – right colic trunk at origin was 4 mm. A CT guided Trucut biopsy revealed histomorphological features suggestive of spindle cell tumour of borderline malignant potential (Fig. 2).

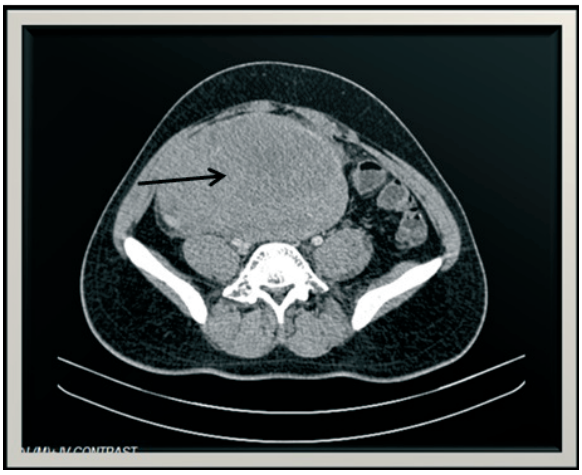

Fig. 1. GIST ** CECT Abdomen and pelvis showing predominantly solid abdominopelvic mass measuring 10×15×16 cm with heterogeneous enhancement. Right sided Grade III hydroureteronephrosis noted.

On exploratory laparotomy there was an intrabdominal mass covering the entire right side of abdomen and right side of pelvis (Fig. 3).

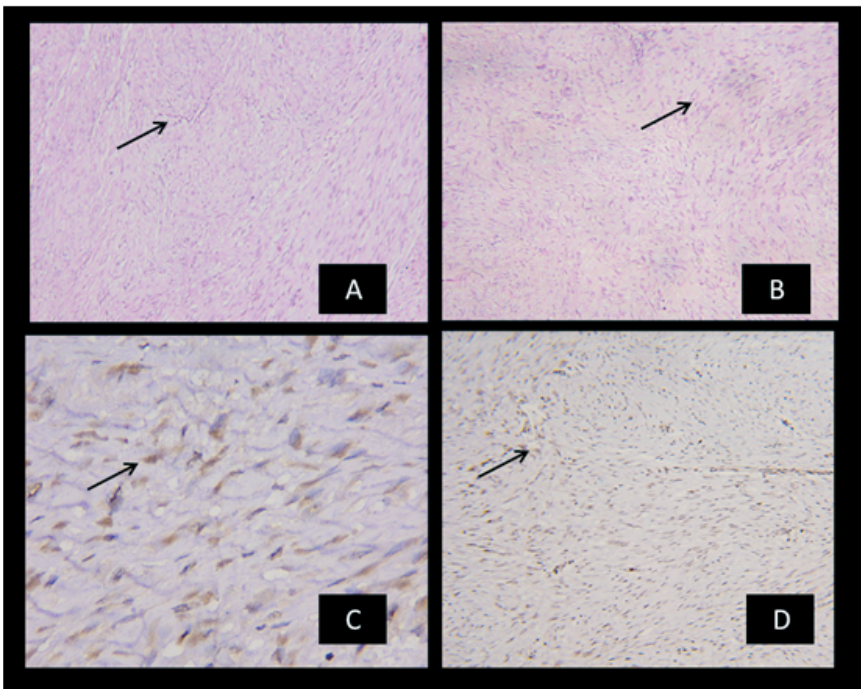

Fig 2. GIST ** A&B: H&E stained section shows interlacing benign spindle cells embedded in myxoid stroma; C&D: Immunohistochemical staining with C kit antibody showing cystoplasmic positivity.

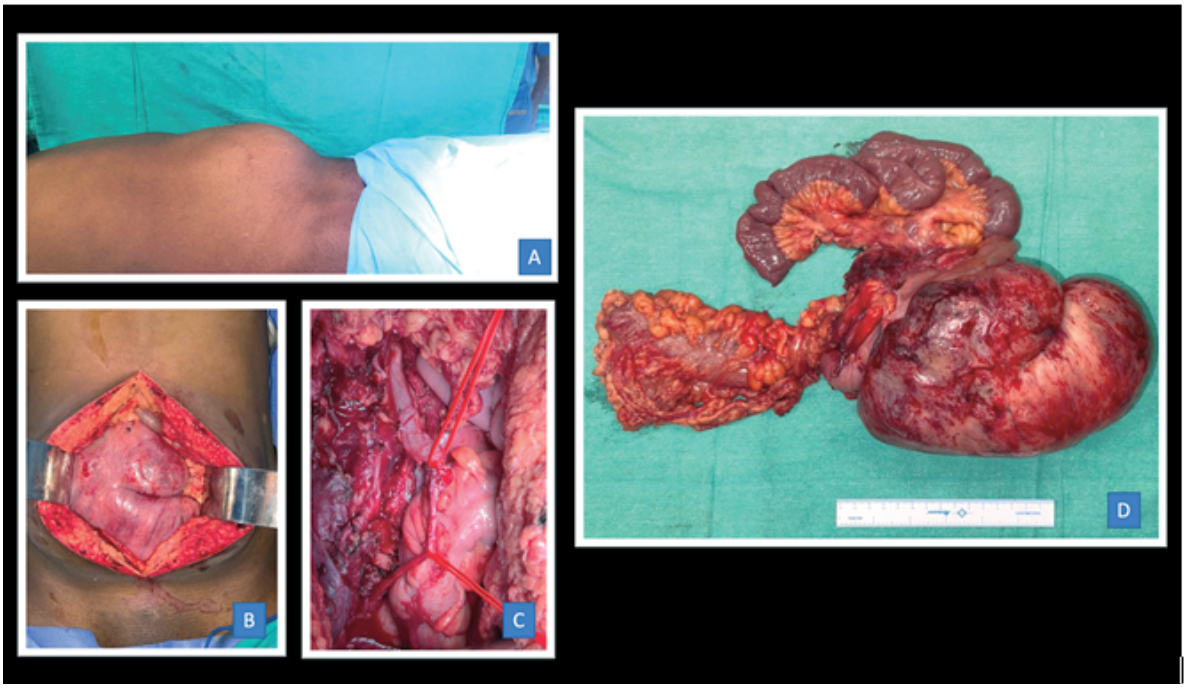

Fig 3. GIST ** A: Preoperational image showing abdominopelvic lump in lateral view; B: Intra pic showing 13×13 cm hard lump; C: Intra op pic showing Right ureter with hydronephrosis; D: Resected specimen showing terminal ileum and transverse colon along with the lump.

The tumour was adherent to the terminal ileum and ascending colon. The tumour was resected with right hemicolectomy and ileo-transverse anastomosis. There was dense adhesion between the retroperitoneum with involvement of right middle ureter. The involved part of right ureter was resected and re-anastomosed with double J stent (Fig. 4).

On the 5th postoperative day, the patient developed increased frequency of stools, which subsided over next few days. He was discharged in 10 days with initiation of chemotherapy with tablet imatinib mesylate 400 mg once daily.

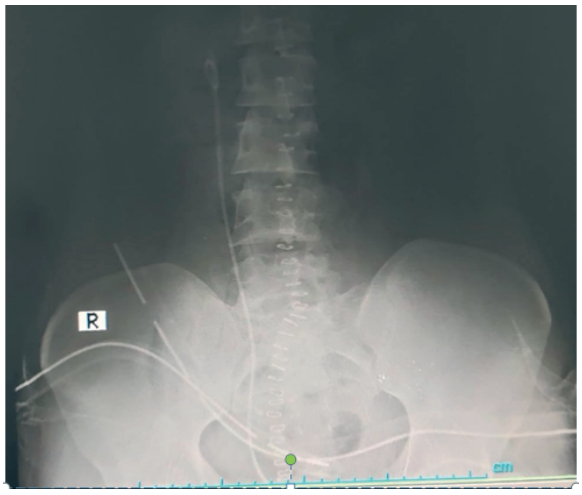

Fig 4. Post op X-ray Pelvis showing Right sided double J stent in place.

The post-operative specimen Histopathology showed Gastrointestinal stromal tumour, spindle cell type, high grade, pT4N0Mx. On immunohistochemistry DOG 1 and CD 17 were positive. CD 34, Desmin, S100 and SMA were negative. Ki 67 score was 8 to 10%. Cut end of the left ureter showed small focus of tumour abutting the adventitia wall. On the follow up in 30 days, the abdominal wall wound healed well and chemotherapy was tolerated without any side effects. DJ stent was removed in six weeks. Review ultrasound examinations over three and sixth month showed no recurrence of growth. Presently the individual has got back to the normal life activities and is perusing a master's course in university.

Discussion

The largest series of retroperitoneal GIST reported in literature comes from Miettinen et al. [1]. They analysed data for a period of 26 years and studied 112 cases of retroperitoneal GIST. Equal incidence was evidenced in both sexes and more common in the elderly population. Avast majority of these tumours were from extraintestinal site with 35 cases from

unspecific sites. All tumours were positive for immunohistochemistry markers for GIST.

Regarding the analysis of the unusual presentation of GIST, many articles are available in the literature. Raja et al. reported a case of GIST presenting as intra peritoneal bleed in a 31-year-old male [2]. The highly malignant nature of the GIST caused ulceration and bleed into the peritoneal cavity and presenting as hemo peritoneum.

Sometimes GIST arising from the retroperitoneum presents with nonspecific symptoms of vomiting, weight loss, low grade fever, anaemia and asthenia. There will be no mass/lump palpable in the abdomen. Thus, imaging technique and biopsy will reveal mass retroperitoneum classified as extraintestinal GIST (E-GIST). Yan et al. reported one of the rare occurrences of pancreatic GIST [3]. This was one of the first two such cases reported in the literature. The diagnosis was made by tissue biopsy using endoscopic ultrasound. A malignant GIST was similarly diagnosed by Rao et al. as a growth in the head of pancreas in a 40-year-old patient [4]. Similarly, Harindhanavudhi et al. [5] reported GIST presenting as haemorrhagic cyst in the head of pancreas.

Another rare location with varied presentation for retroperitoneal GIST is duodenum. The largest series was by Vassos et al. [6]. In their 1-year review of 13 patients this site was amenable to surgical resection. Both local excision and pancreaticoduodenectomy showed comparable results in experience centres. This article also stressed the use of TKI imatinib mesylate in overall long-term prognosis.

Gorelik et al. [7] reported another rare presentation of GIST. The patient presented in bacteraemia with liver abscess. The unusual presentation arose from a fistula between the small intestine and the tumour. Similarly Virgilio et al. [8] studied 15 cases of GIST presenting as mass in the inguinal canal.

Finally, in the review of literature by Gupta [9] 9 cases of gall bladder GIST were reported. These symptoms may be presumed as a sign of mimic cholangitis and are often seen in women. Almost all of these tumours are malignant and require urgent surgical excision.

Finally, a similar case was reported by Ashoor et al. [10] as an incidental finding in a 67-year-old man reported for benign prostrate hyperplasia. The tumour measuring 15 cm was arising from the retroperitoneal aspect of right inguinal fossa. This is the last presentation of incidental findings in radiological evaluation for other problems in this study.

Table 1. Some of the varied presentations of GIST are as tabulated further down

No.	Year	Topic	Author	Presentation
A)	2014	Atypical Presentation of Gastrointestinal Stromal Tumours: A Case Report	Kalpana Raja et al.	Hemangioma
B)	2020	Atypical presentation of gastrointestinal stromal tumor as multiple intra-abdominal cysts: a case report	Ram Prakash Gurram et al.	Gastric cyst
C)	2004	Atypical presentation of gastrointestinal stromal tumor as multiple intra-abdominal cysts: a case report	Ke Kang Sun et al.	Intra-abdominal cyst
D)	2015	Atypical presentation of gastrointestinal stromal tumor masquerading as a large duodenal cyst: a case report	Ameet Kumar et al.	Duodenal cyst
E)	2019	Unusual presentation of a large gist in an extraintestinal site: a challenging diagnosis dilemma	Arwa Ahmed Ashoor et al.	Jejunal mesentry
F)	2018	An Ulcerated Ileal Gastrointestinal Stromal Tumor Disguised as Acute Appendicitis	Ashish Lal Shrestha et al.	Ileal GIST presenting as appendicitis
G)	2017	Ileal gist presenting with bacteremia and liver abscess: a case report and review of literature	Gorelik et al.	Bacteraemia with liver abscess
H)	2021	Inguinal gist: a systematic literature review of primary and metastatic cases	Virgilio et al.	Mass in the inguinal canal
I)	2014	Malignant extra-gastrointestinal stromal tumor of the pancreas: report of two cases and review of the literature	Yan et al.	Pancreatic gist
J)	2019	Gallbladder gist: a review of literature	Gupta et al.	Gall bladder gist

Conclusions

Gastrointestinal stromal tumours are mostly benign neoplasm arising from gastrointestinal tissues. They seldom become malignant and metastasis is rare. There are many atypical variations in clinical presentation for these tumours. The surgeon always needs to keep this diagnosis in mind while evaluating atypical cases of intra-abdominal or retroperitoneal mass.

Limitations

Single centre study.

Conflict of Interests

All authors declare no conflict of interest.

Author's Contributions

Vipin V. Nair, Pawan Sharma, Nagamahendran R. – conceptualization, methodology, formal analysis, writing – original draft, writing – reviewing and editing; *Santosh Raja, Pankaj P. Rao* – data curation, writing – reviewing and editing; *Ritu Mehta* – investigation, formal analysis.

КЛІНІЧНИЙ ВИПАДОК РІДКІСНОГО ЗАОЧЕРЕВИННОГО УТВОРЕННЯ: ТРУДНОЩІ ЛІКУВАННЯ ТА НАБУТИЙ ДОСВІД

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Вступ. *Шлунково-кишкові стромальні пухлини (ШКСП) — це неепітеліальні мезенхімальні солідні новоутворення з різноманітними проявами. У цьому дослідженні описано випадок заочеревинної ШКСП у 21-річного чоловіка, у якого протягом шести місяців спостерігалася припухлість в ділянці живота. Утворення спочатку вважали заочеревинною саркомою. Діагностична лапаротомія виявила черевно-тазову пухлину, що охоплювала всю праву частину живота та таза. Пухлина була спаяна з термінальним відділом клубової та висхідної ободової кишки. Виникли щільні спайки між заочеревинним простором із ураженням середньої третини правого сечоводу. Пухлину видалено з правосторонньою геміколектомією з ілео-поперечним анастомозом. Післяопераційна гістопатологія показала, що це веретеноклітинна ШКСП високого ступеня диференціювання. Зараз пацієнт проходить післяопераційну хіміотерапію іматинібу мезилатом в таблетках.*

Мета. *Атипові прояви ШКСП рідко обговорюються, але часто зустрічаються в клінічній практиці. Ця стаття описує різні проблеми, з якими доводиться зустрічатися хірургу під час діагностики такого нетипового утворення.*

Методи. *Опис випадку атипового ШКСП, який проявлявся у вигляді заочеревинної припухлості.*

Результати. *Пацієнту була проведена хірургічна резекція, зараз він отримує післяопераційну хіміотерапію з хорошим загальним результатом після року спостереження.*

Висновки. *ШКСП, які проявляються у вигляді заочеревинних утворень, зустрічаються дуже рідко, їх слід враховувати при диференціальній діагностиці атипової заочеревинної пухлини.*

КЛЮЧОВІ СЛОВА: ШКСП; атипове положення; заочеревинна пухлина; ущільнення в животі.

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