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Case Report

Radiology Section

A Case of Ewing's Sarcoma of the Jejunum: A Radiological Perspective on Challenges in Diagnosis

TUSHAR KALEKAR[1], M KARTHIK[2], BHAVYA DANG[3], PUSHKAR KUMAR[4]

ABSTRACT

Ewing's Sarcoma (ES) of the jejunum is an exceptionally rare malignancy. It belongs to the family of small round blue cell tumours and primarily affects children and young adults, with gastrointestinal involvement being highly uncommon, particularly in the small intestine. Due to its non specific presentation, including abdominal pain, obstruction, or gastrointestinal bleeding, diagnosis is often delayed, making radiological imaging crucial in differentiating it from other gastrointestinal malignancies such as Gastrointestinal Stromal Tumours (GISTs) and lymphomas. Hereby, the authors present a case report of a 64-year-old male who presented with a three-month history of intermittent abdominal pain, progressive abdominal distension, fever, and loss of appetite. Radiological investigations, including Ultrasonography (USG), Contrast-enhanced Computed Tomography (CECT), and Positron Emission Tomography/Computed Tomography (PET/CT), identified a large circumferential jejunal mass with metastatic involvement of the liver, mesenteric lymph nodes, and peritoneum. Imaging revealed a heterogeneously enhancing mass with central necrosis and high metabolic activity, suggestive of an aggressive neoplasm. Endoscopic biopsy confirmed the diagnosis of ES through histopathological and immunohistochemical markers (CD99, NKX2.2, and synaptophysin positivity). Despite plans for chemotherapy, the patient developed septic shock and multiorgan failure, leading to his death shortly after diagnosis. The present case underscores the critical role of radiological imaging in diagnosing gastrointestinal ES, particularly through CT and Fluorodeoxyglucose-PET (FDG-PET). These imaging modalities help distinguish ES from more common gastrointestinal malignancies, such as GISTs and lymphoma, by identifying characteristic features like heterogeneous masses, necrosis, and metastasis. Early and accurate identification is essential for guiding clinical decision-making and treatment strategies in managing such rare and aggressive tumours.

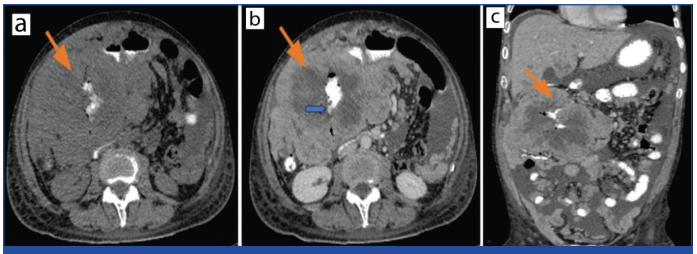
Keywords: Aggressive tumour, Endoscopic biopsy, Heterogeneous mass, Necrosis

CASE REPORT

A 64-year-old male presented with a three-month history of intermittent right lower abdominal pain, which had worsened over the past week, accompanied by abdominal distension, loss of appetite, nausea, and a persistent low-grade fever for two months. The abdominal pain was described as dull and aching, localised to the right lower abdomen, with intermittent acute episodes over the past week. Physical activity, large meals, and specific postures, such as lying on the right-side or bending forward, exacerbated the discomfort. Rest and over-the-counter analgesics, as well as lying on the left-side, provided relief. A recent one-day episode of constipation prompted his visit. He had no significant surgical or medical history.

Examination revealed a pale appearance, stable vital signs, and a non tender, palpable abdominal mass in the right abdomen, which was non contiguous with the liver. Ultrasonography (USG) using a Samsung HS-70A showed a 13×12×12 cm heterogeneously hypoechoic solid mass surrounding a jejunal loop in the right hypochondriac region, adherent to the peritoneum, anterior abdominal wall, and adjacent bowel loops. Dilated proximal small bowel loops suggested obstruction. Enlarged mesenteric lymph nodes with necrotic features and multiple hypoechoic hepatic lesions, likely metastatic, were noted alongside moderate ascites.

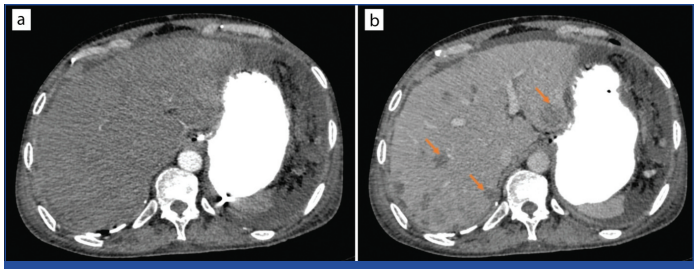
The CECT using a Philips Ingenuity 128-slice scanner confirmed a large, irregularly marginated circumferential jejunal mass (13×12×12 cm) in the right lumbar region with heterogeneous enhancement and central necrotic areas. The mass extended anteroposteriorly from the anterior abdominal wall to the right kidney and cranio-caudally from the liver to the aortic bifurcation, causing a mass effect on adjacent mesentery and bowel loops [Table/Fig-1a-c]. Hypodense non enhancing lesions in both hepatic lobes [Table/Fig-2a,b], mesenteric lymphadenopathy with necrosis, moderate ascites, and generalised subcutaneous oedema were also identified [Table/Fig-3]. The findings indicated a neoplastic jejunal mass with metastatic disease.



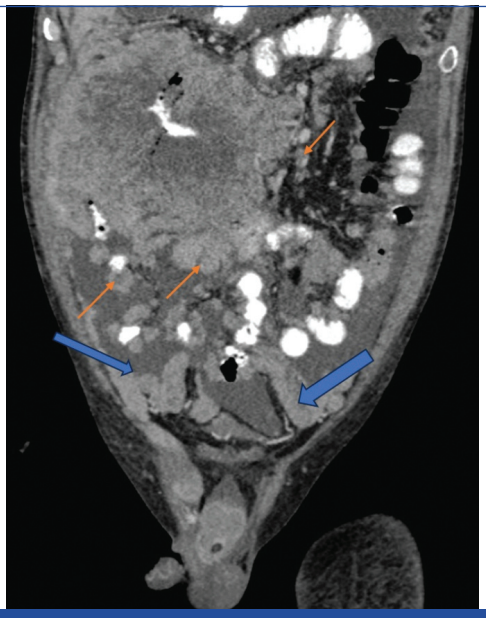
[Table/Fig-1]: a) Non enhanced axial; b) Venous phase image in axial; c) Venous phase image in coronal plane. The above image revealed a large, irregular circumferential mass (orange arrow) in the right lumbar region, surrounding jejunal loop. The mass demonstrated heterogeneous enhancement with central necrotic areas and is causing irregular narrowing of the jejunal lumen (blue arrow).

The FDG-PET revealed intense uptake in the large circumferential jejunal mass, consistent with high metabolic activity [Table/Fig-4], and additional FDG-avid metastatic lesions in the peritoneum, liver, and herniated omentum in the right inguinal canal [Table/Fig-4]. It also confirmed metabolic activity in mesenteric lymph nodes, indicating regional involvement, and identified an FDG-avid thyroid nodule justifying subsequent evaluation. Differential diagnoses established through the above investigations included malignant GISTs and lymphoma, warranting histopathological confirmation.

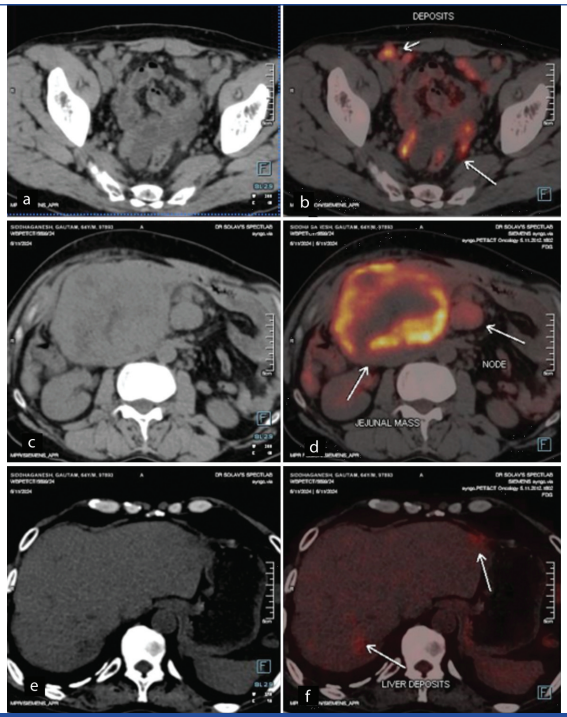
Endoscopic biopsy of the abdominal mass showed sheets and nests of tumour cells with hyperchromatic nuclei, scant cytoplasm, and positive immunohistochemical markers (CD99, NKX2.2, synaptophysin), confirming a peripheral neuroectodermal tumour (ES). Chemotherapy was planned, but the patient developed septic shock and multiorgan failure, ultimately passing away one month later.



[Table/Fig-2]: a) Arterial phase; b) Venous phase. Axial images highlighted multiple small hypodense, non enhancing lesions (orange arrows) in both hepatic lobes, consistent with hepatic metastasis.



[Table/Fig-3]: Coronal venous phase image showed multiple mesenteric lymph nodes including subcentimetric and enlarged nodes (orange arrows), some with necrotic changes, and irregularly enhancing peritoneal deposits (blue arrow).



[Table/Fig-4]: The FDG-PET scan demonstrates intense radiotracer uptake in the large circumferential jejunal mass, reflecting high metabolic activity (c,d). Additional FDG-avid metastatic lesions are observed in the peritoneum, liver, and herniated omentum in the right inguinal canal (a,b,e,f), indicating extensive disease spread.

DISCUSSION

Gastrointestinal Extraskeletal Ewing's Sarcoma (EES) is a rare condition. Primary EES of the small bowel is uncommon; however, it is the most frequent location for gastrointestinal EES [1]. Diagnosing EES in the small bowel is challenging, as the disease often presents with vague and non specific symptoms, making it difficult to differentiate it from other gastrointestinal conditions. As noted in present case, the patient presented with right lower abdominal pain, distension, and systemic symptoms like fever and anorexia. These symptoms are consistent with other cases of gastrointestinal EES noted in the literature [1]. They may resemble many other gastrointestinal conditions, including inflammatory bowel disease or malignant lymphoma. Therefore, radiological imaging plays a crucial role in differentiating EES from these conditions, especially when the clinical presentation overlaps with other gastrointestinal masses, such as GISTs, lymphomas, or metastatic tumours. On imaging, EES presents as heterogeneous masses with necrotic or hemorrhagic areas [1]. In the present case, a CT scan revealed a large irregular mass with heterogeneous enhancement and central necrosis, which are hallmarks of these tumours. This finding is essential for distinguishing EES from other gastrointestinal masses like GISTs, which typically appear more homogeneous on imaging, whereas lymphomas show uniform enhancement and lymphadenopathy.

The Magnetic Resonance Imaging (MRI), although not utilised in present case, is frequently employed as an adjunctive modality, especially for assessing soft tissue involvement and providing more comprehensive imaging of the tumour's extent. MRI findings in EES usually demonstrate high T2-weighted signal intensity, reflecting necrotic or cystic changes, and post-contrast images show significant enhancement, indicative of hypervascularity [2]. These characteristics are essential for surgical or biopsy planning, as they provide improved delineation of the mass and its relationship to adjacent tissues [3].

Furthermore, FDG-PET is essential for staging and identifying metastatic involvement. In present instance, FDG avidity was observed in both the main jejunal mass and the metastatic lesions in the liver and mesenteric lymph nodes, which is characteristic of EES and aids in distinguishing these tumours from benign or less metabolically active masses.

The clinical and imaging findings in present case align with those documented in prior studies of gastrointestinal ES affecting the jejunum. Abdominal pain is commonly noted in patients, with tumour sizes varying from 0.59 to 6.5 cm. Imaging findings are diverse, with certain patients exhibiting homogeneous masses, while others reveal heterogeneous or cystic characteristics [4]. The case displayed a heterogeneous mass, consistent with the imaging features observed in the case report by Cantu C et al., [5].

The ES is defined by histologically uniform small round cells that lack nucleoli, exhibit a finely granular chromatin pattern, and contain minimal cytoplasm, along with varying degrees of neuroectodermal differentiation. The diagnosis of ES relies on a combination of immunohistochemical characteristics, as there is no definitive marker available. The cells exhibit a rounded morphology and show distinct reactions in immunohistochemical assays, including positivity for CD99, vimentin, Friend Leukemia Integration 1 (FLI-1), Leu-7, Erythroblast transformation-specific Regulated Gene (ERG), Neuronal Surface (NS) antigens, and Synaptophysin (SYN) [6,7]. The case results demonstrate positive immunostaining for CD99, NKX2.2, and synaptophysin.

Once ES is diagnosed, management involves a combination of chemotherapy, surgery, and/or radiation. Neoadjuvant chemotherapy, using regimens like VDC/IE (Vincristine, Doxorubicin, and Cyclophosphamide, Ifosfamide Etoposide), aimed to reduce tumour size and address micrometastasis. Surgery is preferred for local control, as it offers better survival outcomes than radiation alone [8,9]. Radiation therapy is also considered when complete resection is not possible.

CONCLUSION(S)

The ES of the jejunum is an extremely rare and aggressive neoplasm characterised by a non specific clinical presentation,

frequently resembling other gastrointestinal disorders, which results in diagnostic delays. The present case underscores the essential role of advanced radiological imaging, including CECT and FDG-PET, in identifying, defining, and staging malignant tumours. A large circumferential jejunal mass exhibiting central necrosis and increased metabolic activity indicated the likelihood of an aggressive tumour, which was later confirmed by histopathological and immunohistochemical analysis. Despite the timely diagnosis and scheduled chemotherapy, rapid disease progression and a lethal outcome highlight the aggressive nature of jejunal ES and the challenges in its treatment. Due to its rarity, more research and clinical knowledge are necessary to improve patient outcomes, enhance therapy strategies, and facilitate early detection.

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PARTICULARS OF CONTRIBUTORS:

1. Professor, Department of Radiodiagnosis, Dr. D. Y. Patil Medical College and Hospital and Research Centre, Pune, Maharashtra, India.
2. Senior Resident, Department of Radiodiagnosis, Dr. D. Y. Patil Medical College and Hospital and Research Centre, Pune, Maharashtra, India.
3. Resident, Department of Radiodiagnosis, Dr. D. Y. Patil Medical College and Hospital and Research Centre, Pune, Maharashtra, India.
4. Resident, Department of Radiodiagnosis, Dr. D. Y. Patil Medical College and Hospital and Research Centre, Pune, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Bhavya Dang,
Resident, Department of Radiodiagnosis, Dr. D. Y. Patil Medical College and Hospital and Research Centre, Pune-411018, Maharashtra, India.
E-mail: bhavyadang1997@gmail.com

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