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Prolonged Fever as an Unusual Presentation of a Mid-Ileal Gastrointestinal Stromal Tumor (GIST): A Rare Case Report

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1. ABSTRACT

Gastrointestinal Stromal Tumors (GISTs) are rare mesenchymal tumors that can develop throughout the gastrointestinal tract, with the majority occurring in the stomach and small intestine. While common presentations include abdominal pain and gastrointestinal bleeding, atypical cases can pose diagnostic challenges. We present a unique case of a 45-year-old man who exhibited prolonged fever as the sole presenting symptom, ultimately diagnosed as a mid-ileal GIST with central ischemic degeneration. Diagnostic workup revealed an 8 cm × 8 cm cavitary lesion on contrast-enhanced CT imaging, confirmed histologically as a spindle cell GIST with low mitotic activity. Surgical resection with end-to-end bowel anastomosis resulted in favourable outcomes. This case highlights the importance of systematic evaluation in pyrexia of unknown origin and emphasizes a multidisciplinary approach to managing atypical GIST presentations.

2. Keywords: Gastrointestinal stromal tumor (GIST); Pyrexia of unknown origin (PUO); Small intestine tumor; Spindle cell GIST; Laparoscopic surgery

3. INTRODUCTION

Gastrointestinal Stromal Tumors (GISTs) are rare mesenchymal tumors, accounting for approximately 0.2% of all gastrointestinal neoplasms. They can develop anywhere along the gastrointestinal tract, with around 50% occurring in the stomach and 30% in the small intestine. The clinical manifestations of GISTs are diverse and influenced by factors such as tumor size, location, and the presence of mucosal ulceration. Common symptoms

include abdominal pain and gastrointestinal bleeding, though less frequently, they may present with intussusception or bowel obstruction [1].

The infrequency of GISTs and their nonspecific clinical features often result in diagnostic delays, emphasizing the need for a high degree of clinical suspicion. Most literature on GISTs focuses on cases involving gastrointestinal bleeding, intussusception, or obstructive symptoms. However, the presented case is unique in its atypical presentation with fever as the sole symptom and the absence of other common clinical features.

4. CASE PRESENTATION

A 45-year-old man, with no prior medical issues, presented with a one-month history of fever occurring intermittently, particularly at night, accompanied by mild weight loss and a reduced appetite. He denied experiencing chronic cough, chest pain, or symptoms related to bowel or urinary function. He had no history of smoking or alcohol use, nor any notable surgical procedures in the past.

On physical examination, there were no remarkable findings, including no significant lymphadenopathy. Respiratory, cardiovascular, and neurological examinations were unremarkable. The abdominal examination revealed slight tenderness in the central region, with no additional abnormalities detected.

Laboratory investigations showed a total white blood cell count of 114.1 × 10^9/L, with neutrophils accounting for 78%. Inflammatory markers were elevated, including a C-reactive protein level of 130 mg/L and an erythrocyte sedimentation rate of 100 mm/hour. Serum lactate dehydrogenase levels were within normal limits, as were renal and liver function tests. Both urine and blood cultures were negative. Peripheral blood smear revealed normochromic normocytic red cells with rouleaux formation and the presence of toxic neutrophils alongside monocytosis. A Mantoux test was also negative.

Radiological investigations included a normal chest X-ray and transthoracic echocardiogram, with no signs of endocarditis on transoesophageal echocardiography. Abdominal ultrasound revealed a segment of thickened bowel and prompted further evaluation with contrast-enhanced CT. The CT scan identified an 8 × 8 cm thick-walled cavitary lesion in the central abdomen, characterized by an air-fluid level and mild fat stranding. The findings suggested a tumor-like lesion, potentially a Gastrointestinal Stromal Tumor (GIST).

A diagnostic laparoscopy was performed, revealing a mass in the mid-ileum with a few enlarged mesenteric lymph nodes. The rest of the abdomen and pelvis appeared normal. Given the lesion's size (approximately 9 cm), a segmental bowel resection was carried out through a small midline incision using a wound retractor, followed by end-to-end anastomosis of the bowel. The patient’s postoperative course was uneventful, and he was discharged on the sixth day after surgery.

Histopathological analysis showed a spindle cell lesion with central ischemic necrosis. Immunohistochemistry revealed the lesion was positive for CD117 and had a Ki-67 proliferation index of less than 1%, while markers such as SMA, Desmin, and CD34 were negative. These findings confirmed the diagnosis of a gastrointestinal stromal tumor with cystic degeneration. The patient was subsequently referred to an oncologist for further treatment.

5. DISCUSSION

Gastrointestinal Stromal Tumors (GISTs) are uncommon neoplasms of the gastrointestinal tract that arise from pluripotent mesenchymal stem cells, which differentiate into interstitial Cajal cells. Approximately 50% of GISTs occur in the stomach, while about 30% are found in the small intestine [1,2]. The clinical manifestation of these tumors varies significantly, depending on their size, location, and the presence of mucosal ulceration. Immunohistochemical analysis typically shows positivity for KIT (CD117). The clinical behaviour of GISTs ranges from benign to malignant, largely determined by their size and mitotic activity. Histologically, GISTs can be categorized into three types: spindle cell, epithelioid, or mixed (spindle and epithelioid). The spindle cell type is the most prevalent, with 95% showing CD117 positivity [3].

GISTs may be asymptomatic and incidentally identified during cross-sectional imaging for unrelated conditions or endoscopic evaluations. Symptomatic tumors often present with nonspecific abdominal pain or discomfort. Tumors larger than 4 cm may cause gastrointestinal bleeding [2], and in rare cases, they may lead to complications such as intussusception or bowel obstruction. In this case, while the patient experienced mild nonspecific pain, the primary concern was a prolonged fever lasting nearly six weeks, qualifying as Pyrexia of Unknown Origin (PUO). A thorough evaluation with relevant investigations was essential to establish a diagnosis. This type of presentation is exceedingly rare and scarcely documented in surgical literature.

Contrast-enhanced CT imaging is particularly useful for identifying small bowel GISTs [4]. Once diagnosed, the primary treatment is surgical resection, aiming to remove the tumor with adequate margins while preserving the intact pseudo capsule. GISTs rarely metastasize to regional lymph nodes, making lymphadenectomy unnecessary [5]. The tumor's behaviour and the need for adjuvant therapy with imatinib, an oral kinase inhibitor, depend on the histological features of the tumor [5]. In this case, the tumor was of spindle cell type, positive for CD117, with a low Ki-67 proliferative index, indicating a low-risk lesion.

This case represents an exceptionally rare presentation of a mid-ileal GIST, where the patient exhibited prolonged fever attributed to central ischemic degeneration of a large tumor measuring nearly 9 cm. Systematic evaluation and thorough investigations are crucial for diagnosing PUO. Careful planning and the use of a laparoscopic approach resulted in a favourable surgical outcome. This case underscores the importance of a multidisciplinary approach in patient management, with valuable contributions from radiology and pathology, including immunohistochemical analysis.

6. CONCLUSION

In conclusion, this case highlights the rare and atypical presentation of a Gastrointestinal Stromal Tumor (GIST) as Pyrexia of Unknown Origin (PUO), which led to a diagnostic challenge. The patient's prolonged fever, coupled with nonspecific abdominal symptoms, made the diagnosis difficult, emphasizing the need for a high index of suspicion in such cases. Despite the rarity of this presentation, the combination of imaging, diagnostic laparoscopy, and histopathological analysis led to the accurate diagnosis of a mid-ileal GIST. The patient's successful outcome following surgical resection further underscores the importance of timely diagnosis and the role of a multidisciplinary team in managing complex cases. This case contributes to the limited literature on GISTs presenting with fever and serves as a reminder of the diverse clinical manifestations these tumors can exhibit.

7. REFERENCES

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