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Case Report

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Chronic myeloid leukaemia presenting as acute small bowel gangrene: A case report

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ABSTRACT

Rationale: Chronic myeloid leukaemia is a myeloproliferative disorder due to clonal hyperproliferation of myeloid cells within the bone marrow. It can present both pro- and anti-thrombotic states. CML has different presentations within the gastrointestinal tract.

Patient's concern: A 40-year-old non-diabetic and non-hypertensive male complained of abdominal pain with nausea and emesis for 1 day. Besides, he had a history of abdominal distension and fever for 1 day.

Diagnosis: Acute small bowel gangrene due to chronic myeloid leukaemia.

Intervention: A limited resection of small intestine with ileostomy and mucus fistula.

Outcome: After 3 months following surgery the patient underwent stoma closure. The patient was followed up for more than 3 years postoperatively. During the follow-up, the patient was asymptomatic without any recurrence of the disease.

Lesson: Chronic myeloid leukaemia should be considered as one of the causes for small intestine gangrene when there is increased leukocyte count, splenomegaly without evidence of atherosclerotic occlusion or systemic emboli from the heart.

KEYWORDS: Chronic myeloid leukaemia; Superior mesenteric artery thrombosis; Acute small bowel gangrene; Myeloproliferative disorder; Prothrombotic state

1. Introduction

Chronic myeloid leukaemia (CML) is a myeloproliferative disorder due to clonal hyperproliferation of myeloid cells within the bone marrow. It is characterized by a reciprocal translocation between chromosomes 9 and 22. In the myeloproliferative disorder, bleeding manifestation and thrombotic manifestations can be found. Thrombotic manifestations are less common in CML compared to other myeloproliferative disorders like polycythaemia vera and essential thrombocytosis. This study reported a rare case of CML presenting as acute small bowel gangrene due to superior mesenteric artery thrombosis.

2. Case report

This study was approved by the Ethical Committee of the Institutional Ethics Committee (human studies), SRM Institute, and informed consent was obtained from the patient.

A 40-year-old non-diabetic and non-hypertensive male complained of abdominal pain with nausea, emesis, and constipation for 1 day. Besides, the patient reported a history of abdominal distension and fever for 1 day. There was no previous history of surgery or medical co-morbidities. On examination, the patient became dehydrated. The blood pressure was 100/60 mmHg and pulse rate was 104/min. Per abdominal examination shows abdominal distension with splenomegaly. Non-rigidity abdomen with voluntary guarding was noted. There was no significant abnormality in the cardiovascular examination and the respiratory tract examination.

The preoperative blood investigations showed anemia with increased white blood cells of 36000 cells (leucocytosis) (normal range: 4000-11000). The renal function test and the liver function

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test were normal. The patient underwent an erect abdominal X-ray showing dilated small bowel loop without pneumoperitoneum (Figure 1). Besides, contrast computed tomography of the abdomen showed decreased contrast enhancement in the ileum with massive splenomegaly and splenic infarct (Figure 2). A pre-operative echocardiogram was done as part of the screening, which showed no regional wall motion abnormality and no evidence of thrombus.

The patient was diagnosed with acute small bowel gangrene based on the clinical and radiological findings and underwent diagnostic laparoscopy, which showed 15 cm of gangrenous ileum (Figure 3). So, we converted laparoscopy into laparotomy. On further exploration, we were able to identify gangrenous ileum along with thrombus in the superior mesenteric artery (Figure 4). In the view of thrombus in the superior mesenteric artery, we did limited resection of the small intestine with ileostomy and mucus fistula. Post-operatively, the patient was on anticoagulant (injection of low molecular weight heparin). Postoperative histopathological examination of the resected small intestine showed necrotic small bowel loops. Since there was no evidence of thrombus in the heart, we wanted to identify the cause for superior mesenteric artery thrombosis. So, we did a peripheral smear, which showed immature granulocyte with eosinophilia and basophilia. Besides, neutrophilia and monocytosis were found as well. The bone marrow study showed increased granulocyte precursors, basophils, eosinophils, and monocytes. There were increased megakaryocytes and myeloid cells. The cytogenetics of the patient showed the Philadelphia chromosome with *ABL* gene and *BCR* gene translocation; t (9;22) (q34;q11). The patient was started on an anti-tyrosine kinase inhibitor (Imatinib) 400 mg once a day in the post-operative period.

The patient was followed up for more than 3 years since surgery. During follow-up, the patient is asymptomatic without any recurrence of the disease. After 3 months following surgery, the patient underwent stoma closure.

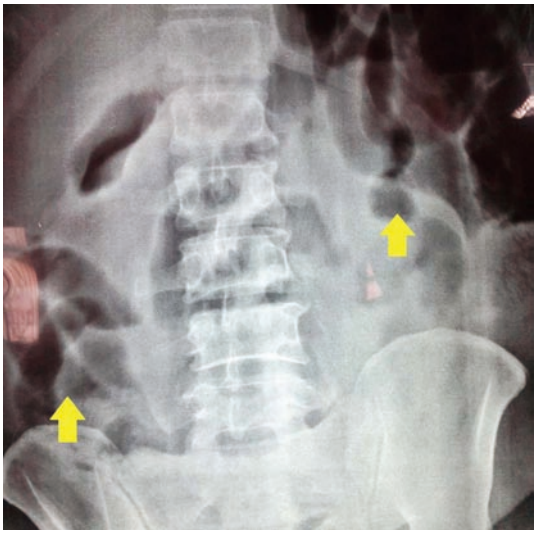

Figure 1. Abdomen X-ray shows dilated small bowel loops (arrows).

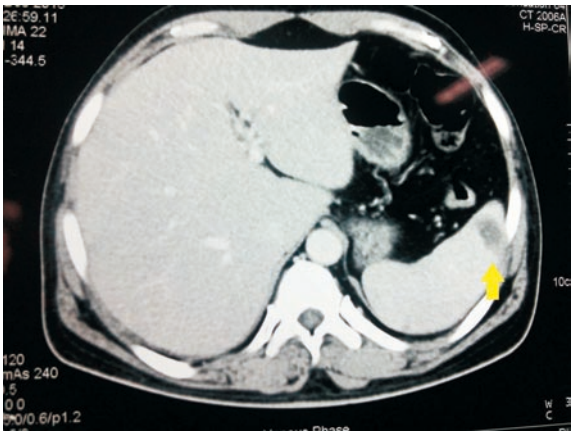

Figure 2. CT axial view shows splenomegaly along with splenic infarct (arrow).

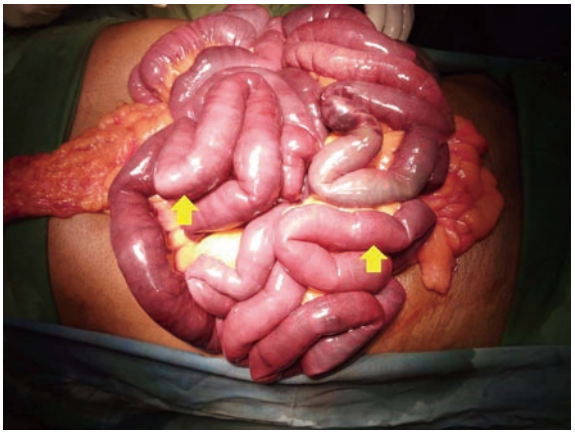

Figure 3. The intraoperative photograph shows dilated small bowel with gangrene (arrow).

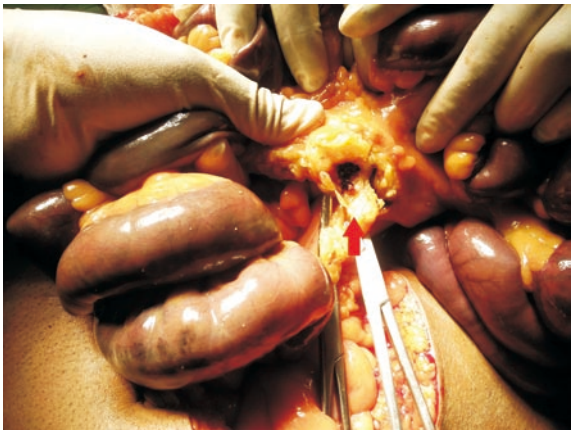

Figure 4. The intraoperative photograph shows a thrombus in the superior mesenteric artery (arrow).

3. Discussion

CML is a myeloproliferative disorder due to clonal hyperproliferation of myeloid cells within the bone marrow[1]. The presentations of CML vary from anorexia, loss of weight, loss of appetite, fever, *etc.* It also presents with features of thrombosis like avascular necrosis of the femur, ophthalmic vein thrombosis, splenic infarct, *etc*.

The hematopoietic cells contain reciprocal translocation between chromosomes 9 and 22[1]. The acquisition of the BCR-ABL fusion is a result of translocation of t (9,22) (q34;q11.2) in a single multipotential hematopoietic cells[2]. Both bleeding and thrombosis occur in myeloproliferative disorders.

The thrombotic events are less common in CML compared to other myeloproliferative disorders like polycythaemia vera and essential thrombocytosis. There are reports of splenic infarct[2], portal vein thrombosis, intussusception in the gastrointestinal tract following CML, but superior mesenteric artery thrombosis was rarely reported[3]. The gastrointestinal manifestations of leukaemia are due to leukemic cell infiltration and vascular complications because of the bleeding and thrombosis[3].

Leukaemia can affect the entire gastrointestinal tract starting from the oesophagus up to the large intestine[4]. In the small and large intestine, it can present as lower gastrointestinal hemorrhage, colitis, intussusception, obstruction, ulceration, perforation[5]. Apart from the above complications, secondary gastrointestinal malignancy like colon cancer is also reported with chronic myeloid leukaemia.

Abdomen X-ray is a non-specific diagnostic tool, and mostly it shows dilated bowel loops. Ultrasound examination of the abdomen might reveal organomegaly or solid organ infarct with lymphadenopathy. Contrast-enhanced computed tomography is an investigation to identify vascular occlusion and necrosis of bowel loops. The role of endoscopy is limited in the diagnosis of gastrointestinal manifestations of chronic myeloid leukaemia.

The treatment of CML purely depends on the organ involvement and stage of the disease. Most of the cases with ischemic gastrointestinal disease required resection of the involved segment followed by anastomosis[6]. All the patients need post-operative anticoagulants along with primary treatment for chronic myeloid leukaemia[7].

In conclusion, chronic myeloid leukaemia should be considered one of the causes for acute small intestine gangrene, when there is increased leukocyte count, splenomegaly without evidence of atherosclerotic occlusion or systemic emboli from the heart.

Conflict of interest statement

The authors report no conflict of interest.

Authors' contributions

J.P: Concept and design of the study, acquisition of data, drafting the article; J.P, A.S: Acquisition of data revising the article critically for important intellectual content.

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