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GI IMAGE

Small Bowel Volvulus Caused by Small Intestinal B Cell Lymphoma

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Case Presentation

A 67-year-old otherwise healthy male with a remote history of trauma and right perinephric hematoma presented to the hospital for acute onset severe left-sided abdominal pain, fever of 39 °C, inability to tolerate diet, and 12 lbs of weight loss over 1 month. On physical exam, he had tachycardia and significant left-sided abdominal tenderness. The laboratory studies were notable for leukocytosis of 16,200 μl and hemoglobin of 8.5 g/dl which dropped to 6.7 g/dl after resuscitation. He underwent a CT abdomen which showed a large dilated thickened loop of bowel along with retroperitoneal lymphadenopathy concerning for lymphoma (Fig. 1).

Additionally, he was noted to have a subcapsular right renal collection most likely related to previous trauma. Due to fevers, significant abdominal pain, thickened bowel loop, and acute on chronic anemia, he was taken to the operating room for exploratory laparotomy. At surgery, an approximately 25-cm mass which was adherent but not invading the abdominal wall was noted (Fig. 2). The mass and the small intestine were eviscerated. The mass appeared to be a thickened loop of the terminal ileum. There was an additional full thickness diverticulum between the mass and the ileocecal valve. There was also an internal hernia caused by adhesion between the appendix and the mass. The mass, diverticulum, and adhesive appendiceal band were all removed en bloc with right colectomy and primary anastomosis. The patient was discharged home on postoperative day two on oral antibiotics.

The pathology of the resected specimen showed a 25 cm × 17.5 cm mass composed of diffused atypical lymphoid infiltrates stating positive for CD20 and BCL2 and negative for other markers consistent with B cell lymphoma. Additional hematopathology consultation revealed a diagnosis of small B cell lymphoid neoplasm with plasmacytoid differentiation, the differential diagnosis being extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT) or lymphoplasmacytic lymphoma. The bone marrow biopsy did not show any evidence of involvement by lymphoma supporting a diagnosis of MALT lymphoma. On PET/CT, metabolically active lymphadenopathy was seen in the mediastinum, retroperitoneum, and pelvis. Additionally, the perinephric infiltration was also metabolically active suggestive of diffused infiltration by lymphoma. The patient is currently undergoing chemotherapy with rituximab and bendamustine.

Discussion

Although lymphoma is not a surgical disease, surgery may have a role in some patients with primary gastrointestinal (GI) lymphomas who present with acute abdomen. Primary GI lymphomas comprise 5–20% of extra-nodal lymphomas making them the most common type of extra-nodal disease.[1] Age and sex-adjusted yearly incidence rates of primary GI lymphomas ranged from 0.13 per 100,000 in 1999 to 2.39 per 100,000 in 2007.[2] The most common distribution of disease is duodenum (65%), followed by jejunum and ileum (20%).[3] The majority of primary GI lymphomas are non-Hodgkin's lymphomas

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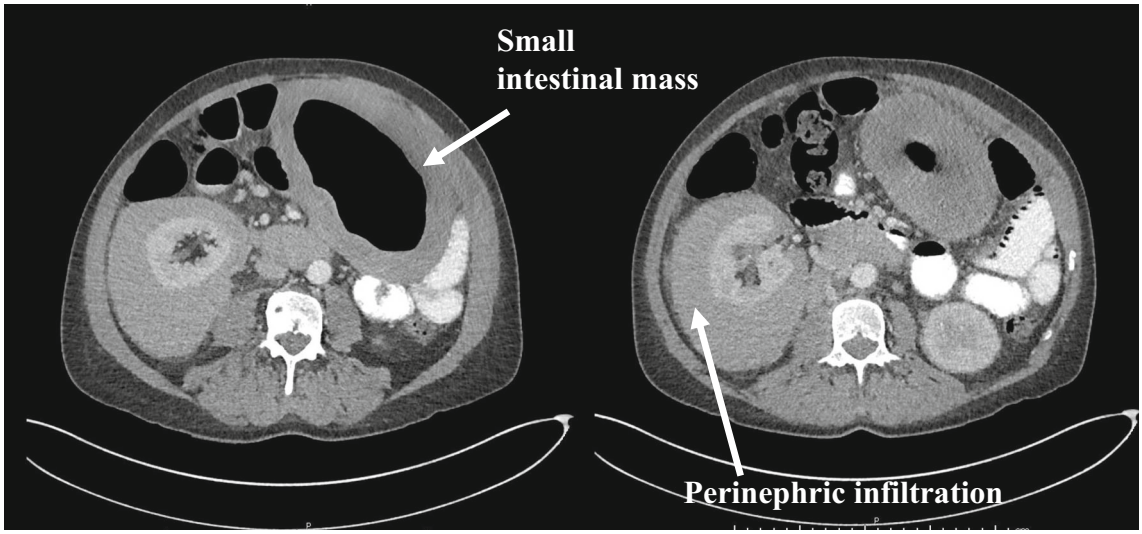

Fig. 1 Large dilated thickened loop of bowel along with retroperitoneal lymphadenopathy and a right subcapsular renal collection concerning for lymphoma

with diffused large B cell lymphoma (DLBCL) being the most common subtype (47%).[2] These are often associated with poor prognosis. Mucosa-associated lymphoid tissue (MALT) lymphomas are the next most common (24%) and are associated with chronic inflammatory states such as Sjögren syndrome or *Helicobacter pylori* infections.[1,2] Chronic inflammation leads to local accumulation and proliferation of antigen-dependent B cells and T cells.[1] Less common are follicular lymphomas and mantle cell lymphomas which are driven by overexpression of BCL2 and cyclin D1, respectively.[2,3] Surgical resection is often the initial mode of treatment for primary small intestinal lymphomas due to difficulty in obtaining a pre-operative diagnosis.[1]

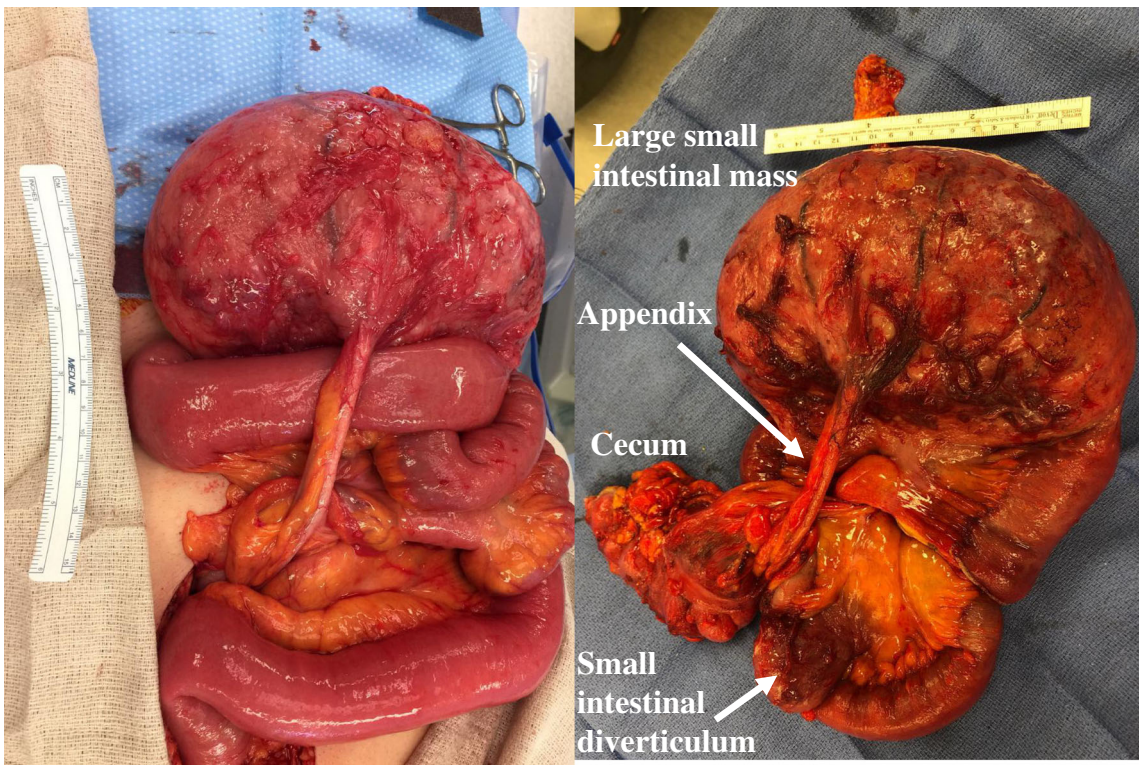

Fig. 2 An approximately 25-cm mass which was adherent but not invading the abdominal wall. The mass was small intestinal in origin, but was tethered to the appendix, causing volvulus symptoms

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflicts of interest.

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