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ADULT ILEOILEAL INTUSSUSCEPTION CAUSED BY DIFFUSE LARGE B-CELL LYMPHOMA

Scarlet Louis-Jean
Anne Arundel Medical Center Inc, scarletl@auamed.net

Shire Chaudhry

Stephanie Richards

Adult Ileoileal Intussusception Caused by Diffuse Large B-cell Lymphoma☆

Scarlet Louis-Jean*, Shire Chaudhry, Stephanie Richards

Internal Medicine Department, Luminis Health Anne Arundel Medical Center, Annapolis, MD, USA

Abstract

Adult intussusception is a rare diagnosis that can be caused by non-Hodgkin's diffuse large B-cell lymphoma (DLBCL). In this case report, we discuss a middle-aged man who presented with non-specific symptoms of intussusception and absence of classic B symptoms. He was found to have intussusception secondary to stage IIIE, CD20 positive DLBCL. The patient underwent small bowel resection with anastomosis, followed by 6 cycles of R−CHOP, which resulted in complete remission of his neoplasm. In reporting this case, we hope to further highlight the role of malignancy in intussusception and guidance on appropriate therapy.

Keywords: Diffuse large B cell lymphoma, Intussusception, Non-hodgkin's lymphoma, R−CHOP

1. Introduction

Intestinal intussusception, or intestinal telescoping into a more distal bowel segment, is a rare cause of mechanical bowel obstruction, accounting for 1−5% of cases in adults.[1-3] A lead point is often the underlying cause of intussusception. This could be a benign or malignant mass, such as in lymphoma. The gastrointestinal tract is the most common extra nodal site affected by lymphoma (5−20%).[1] Histopathologically, almost 90% of primary gastrointestinal lymphomas are B-cell non-Hodgkin's lymphomas (NHL) with the greatest involvement seen in the stomach (50−60%), small intestine (20−30%), and colorectum (10−20%).[1,3,4] While intussusception is a very rare presentation of NHL, the most common lymphoma causing intussusception is DLBCL.[1]

2. Case report

A 46-year-old Black man with no previous medical history presented to the emergency department with intermittent diffuse abdominal pain for one month, which worsened in severity and in persistence, the week of arrival to the emergency department. The week prior to admission, the patient described inappetence, nausea, vomiting, and constipation, without fever, night sweats, unintentional weight loss, or bloody stools. Laboratories were unremarkable. He was found to have mild generalized tenderness without peritonitis on abdominal palpation. CT abdomen with intravenous contrast revealed obstructing intussusception of the distal ileum, proximal to the terminus (Fig. 1). The patient underwent exploratory laparotomy for small bowel resection of a 4.9 cm transmural mass,

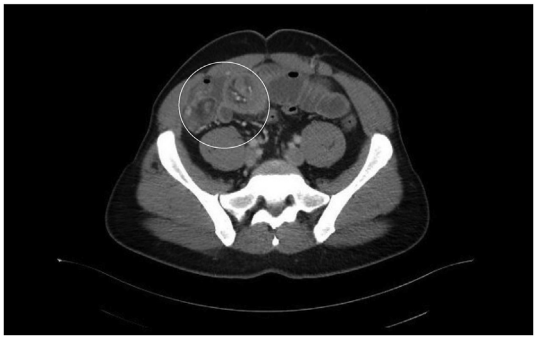

Fig. 1. CT abdomen and pelvis without contrast showing an area of intussusception (circle) within the mid to distal small bowel.

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* Corresponding author.
E-mail address: ScarletL@auamed.net (S. Louis-Jean).

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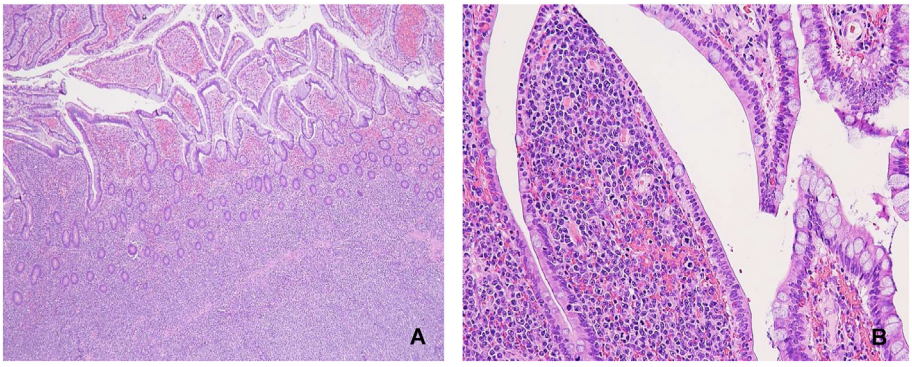

Fig. 2. (A) Low power (20X) hematoxylin and eosin (H&E) image showing an atypical lymphoid infiltrate involving the small bowel mucosa, submucosa, and muscularis propria. (B) On higher power (100X), the atypical infiltrate consists of large lymphoid cells with prominent nucleoli and increased mitotic activity, morphologically consistent with diffuse large B cell lymphoma (DLBCL, left side). Normal small bowel villi are present on the right side for comparison.

followed by enteroenterostomy. Surrounding mesenteric lymph nodes were also excised.

Surgical pathology of the small bowel resection revealed stage IIIE, CD20 positive atypical lymphoid infiltrates involving the small bowel mucosa, submucosa, and muscularis propria, comprising of large lymphoid cells with prominent nucleoli and increased mitotic activity, consistent with diffuse

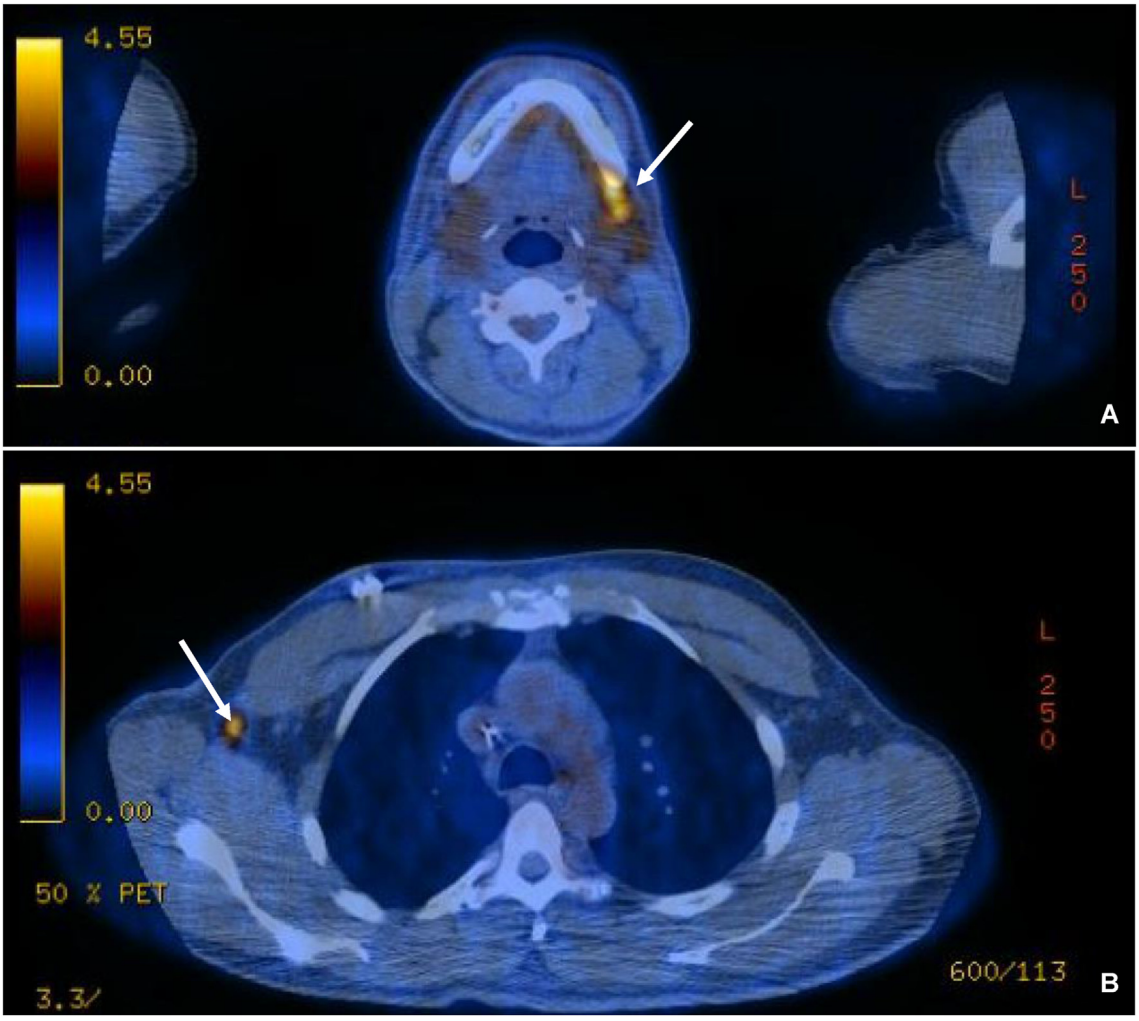

Fig. 3. PET CT showing clustered hypermetabolic level 1B left perimandibular nodes (arrow) consistent with a localized lymphoproliferative disorder or benign reactive nodal change (A), and a nonspecific hypermetabolic right axillary lymph node indicated by the arrow (B).

large B cell lymphoma (Fig. 2). High grade fluorescence in situ hybridization (FISH) analysis demonstrated absence of MYC, BCL2, or BCL6 rearrangement, suggesting an absence of double/triple hit lymphoma (DHL/THL). Mesenteric lymph node biopsy showed fragments of lymph nodes involved by DLBCL, which were strongly positive for MUM1 and BCL2, with high Ki-67 expression. PET scan indicated clustered hypermetabolic level 1B left perimandibular nodes in addition to hypermetabolic right axillary lymph nodes (Fig. 3). The patient underwent 6 cycles of chemotherapy with Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone (R–CHOP) every 21 days. Follow up PET scan performed within a month of treatment completion showed no evidence of abnormal PET avidity, suggesting complete response to therapy.

3. Discussion

Adult intussusception is a rare occurrence, often resultant of an imbalance in longitudinal forces caused by a mass acting as a lead point, in association with disorganized peristalsis.[1] The most common lead points are benign, while malignant tumors account for 30% of presentations.[1,2] There are 3 major subtypes of adult intussusception, which include: entero-enteric, ileo-cecal, and colo-colic.[1] Unlike pediatric cases, adult intussusception can often be associated with a palpable abdominal mass 9–63% of the time.[1] The common presenting symptoms of intussusception are non-specific and include abdominal colic, nausea, vomiting, bleeding, diarrhea, and constipation.[1] The diagnosis is often made with CT, which has a sensitivity of 58–100%.[3] The treatment of choice is surgical resection, and in cases caused by malignancy, appropriate staging with tissue biopsy and PET/CT is indicated prior to chemotherapy.

DLBCL is a common heterogeneous subtype of NHL, characterized by monoclonal proliferation and expansion of large and mature B-cells that arise from primary, *de novo*, or transformation of an indolent lymphoma.[5,6] It represents 25–31% of yearly NHL cases in western countries.[6,7] Staging is determined following tissue biopsy using the Lugano criteria, which defines early-stage disease as stage I or II, and advanced stage disease as III or IV.[6,7] Epidemiologically, DLBCL occurs predominantly in males in their sixth decade.[6] While DLBCL is considered aggressive, it has a high cure rate, as patients with limited disease experience an 80–85% five-year progression free survival (PFS), while those with advanced disease have a 50% five-year PFS.[7]

The standard treatment of DLBCL is chemotherapy with R–CHOP despite disease severity, although treatment tailoring is prognosis-dependent and determined by the cell of origin, revised International Prognostic Index (R–IPI), and age adjusted IPI (aaIPI).[7,8] Prognosis is also dependent on cytogenetic alterations and protein expression, with patients, like the one discussed in this case report, having a poor 5-year outcome survival with expression of MUM1.[8,9] Additionally, BCL2 (apoptosis regulator) and Ki-67 (proliferation marker) expression have been shown to confer poor PFS in DLBCL.[8,9] Nonetheless, patients with DLBCL are often cured with 6–8 cycles of R–CHOP, while 10–15% and 20–30% experience refractory disease and relapse, respectively.[10] Most patients with the poorest outcomes and resistance to chemotherapy often express DHL/THL rearrangements, which our patient did not, as MYC rearrangement is required to meet criteria for either distinction despite his atypical, although clinically insignificant BCL2 and BCL6 activity.[8]

Our patient presented without classical B symptoms, which are often associated with lymphoma, but rather with manifestations of intussusception. Incidentally, he was found to have DLBCL, which was confirmed via tissue biopsy. Altogether, he had a good prognosis score of 2, a 79% overall survival, and 80% PFS, determined by the R–IPI, despite his MUM1, BCL2, and Ki-67 expressions. He achieved total remission following 6-cycles of R–CHOP and is scheduled with hematology/oncology for interval follow-ups to assess for recurrence.

4. Conclusion

Owing to the rarity of intussusception in adults, having a high index of suspicion may be difficult as the presentation is nonspecific and can align with other common abdominal pathologies among adults. CT A/P is the most sensitive modality to help rule out this condition. In adult intussusception associated with malignancy, surgical resection and chemotherapy are the treatments of choice, following appropriate staging with tissue biopsy and PET/CT. In patients with DLBCL, R–CHOP chemotherapy has demonstrated a high cure rate and is the standard of treatment despite disease aggressiveness.

Author contribution

All three authors made substantial contributions to the conception and acquisition of the data for the manuscript. Dr. Chaudhry drafted the initial introduction and case report. Dr. Louis-Jean revised and added substantial additions including the discussion,

and figures and legends. Dr. Stephanie Richards contributed to the synthesis and interpretation of the pathology results of the biopsied mass. All authors agreed with the final version of this manuscript and its submission. Additionally, we agreed that we would be accountable for all aspects of the manuscript.

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Informed consent

Informed consent was obtained from the patient for the publication of the case details.

Conflict of interest

The authors of this manuscript do not have competing interests to disclose.

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