International Journal of Surgery Case Reports 90 (2022) 106748

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

A rare case of primary non-metastatic Non-Hodgkin's diffuse large B-cell lymphoma in the ileum in a 19-year-old male manifested as intestinal obstruction- A case report

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ARTICLE INFO

Keywords:
Case report
Intestinal lymphoma
Primary Ileal Non-Hodgkin's lymphoma
Diffuse large B-cell lymphoma
Intestinal obstruction
Surgical abdomen

ABSTRACT

Introduction and importance: The gastrointestinal system is the most common site for extra-nodal NHL. Adolescent population are among the rarest of compromised groups, especially males. The gastrointestinal system is more involved by secondary metastasis rather than by primary lymphomas. Cardinal B-Symptoms and obstipation constituted the presentation of our patient who was diagnosed postoperatively as a case primary non-metastatic NHL. NHL can have misleading presentations which result in the implementation of different treatment modalities. We ought to have high clinical suspicion when presented with a patient suffering from B-Symptoms and obstipation to make timely judgements which help in performing effective therapeutic interventions to limit the morbidity and mortality which result from this pathology.

Case presentation: We present the case of a 19-year-old male, who presented with obstipation and B-Symptoms. CT scan indicated loop dilation, a lobulated mass, and what radiologically seemed to be intussusception. Surgery was done and the resected specimens were DLBCL.

Clinical discussion: We treated him by surgical excision of the affected ileal segments. Histopathology indicated a primary Non-Hodgkin's DLBCL of the ileum. Afterwards, we referred him for adjuvant chemotherapy. Treatment modalities for this malignancy are mainly surgical in addition to Chemotherapy.

Conclusion: Intestinal extranodal NHL presents with an array of vague symptoms. As a result, this type of tumors can be clinically indistinguishable from other gastrointestinal malignancies. It is vital to keep this type of malignancy in mind as a differential diagnosis when presented with a surgical abdomen in a patient with B-Symptoms.

1. Introduction

Lymphoma can be classified depending on site of occurrence to either nodal or extranodal. The most common system involved in extranodal lymphoma is the gastrointestinal system [1–2]. In descending order, the most prevalent gastrointestinal system location for primary lymphoma is the stomach 50–60%, followed by the small intestine (20–30%) and lastly, the colon and rectum which comprise 10–20% [3–4]. Primary small intestinal lymphoma is extremely rare, it accounts for 0.2% to 0.65% of all colonic neoplasms [5]. Primary gastrointestinal Non-Hodgkin's Lymphoma is responsible for 30–40% of all extranodal Non-Hodgkin's Lymphoma [6] High-grade lymphoma such as Diffuse Large B-Cell Lymphoma typically manifests with ulcers, perforation, and several segments of stenoses [7–8] It presents with a variety of non-specific symptoms and its treatment either involves the sole use of chemotherapy or the combination of surgery and chemotherapy [9].

The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines [10].

Abbreviations: DLBCL, Diffuse Large B-Cell Lymphoma; NHL, Non-Hodgkin's Lymphoma; CT, Computed Tomography; ED, Emergency Department; H&E, Hematoxylin and Eosin; R-CHOP, Rituximab- Cyclophosphamide- Hydroxydaunorubicin- Oncovin- Prednisone.

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https://doi.org/10.1016/j.ijscr.2021.106748
Received 19 November 2021; Received in revised form 8 December 2021; Accepted 31 December 2021
Available online 4 January 2022

2. Presentation of case

2.1. Patient information

We present the case of a previously healthy, 19-year-old Middle Eastern male who presented as a case of acute surgical abdomen. The story started 2 weeks prior to admission with the patient complaining of non-specific abdominal pain which was intermittent and ill-defined in nature. In addition, he complained of unprecedented loss of appetite, nausea, and undocumented weight loss, night sweats, and bowel habits alternating between constipation and diarrhea. The pain became peri-umbilical and migrated to the right iliac fossa 5 days prior to admission and as a result, he was taken to the Emergency Room in a suburban hospital and thus, was diagnosed with acute Appendicitis and was treated with an Appendectomy. He was discharged home on the following day. However, his symptoms did not subside postoperatively and as a result, he presented to our Emergency Department via ambulance with a 3-day-history of fatigue, obstipation and colicky peri-umbilical and right iliac fossa abdominal pain accompanied by nausea, multiple episodes of non-biliary vomitus, and loss of appetite. The pain was unresponsive to over-the-counter analgesics. Undocumented fever was reported. Nonetheless, no genitourinary symptoms were reported. Negative family, drug, and allergic histories. He doesn't smoke nor consumes alcohol. His BMI is 23 Kg/m^2.

2.2. Clinical findings

Clinical examination revealed tachycardia and tachypnea. Otherwise, vital signs were normal. Upon inspection, the abdomen was symmetrical, but painfully moves with respiration. A McBurney's scar incision was noted. On palpation, there was peri-umbilical guarding and tenderness. Bowel sounds were roughly auscultated. Laboratory investigations revealed leukocytosis (WBC: 10.5/μL) and hypokalemia (Potassium: 3.2 mmol/L), but otherwise, within normal values.

2.3. Diagnostic assessment

Abdominal Ultrasound revealed normal Liver and Spleen. Additionally, significant dilation of small bowel loops was noted. However, the rest could not be studied due to the dilation of bowel loops. CT scan of the Abdomen and Pelvis suggested thickening of the small bowel loops in the right iliac fossa in addition to what appears to be an invagination of the small bowel mesentery. Moreover, there is ileal and jejunal loops dilation and an ileal transitional zone causing stenosis proximal to the ileocecal valve. Findings are vaguely suggestive of intussusception of the ileum (Fig. 1A–B) -as indicated by the circle and the arrow- A mass-like structure with lobulated borders situated anterio-inferior to the left kidney compressing the small bowel loops proximal to the ileocecal valve (Fig. 1C) -as indicated by the arrows- These findings can explain the obstructive symptoms of the patient.

Preliminary therapy involved intravenous fluid resuscitation, analgesics, prophylactic antibiotics, full spectrum laboratory testing including sampling and crossmatch of blood group.

The challenges faced were the unavailability of a laparoscopic device, an interventional radiological device in the Emergency Department at that time, and the low socioeconomic status of the patient which limited the ability to stain for all the possible immunohistochemical stains for DLBCL to only the most essential ones.

2.4. Therapeutic intervention

An Exploratory laparotomy was indicated depending on the clinical examination of the patient and because the radiological analysis suggested intestinal obstruction. The operation was performed at our tertiary university hospital. It was done by two senior year fifth year General Surgery residents with five years of experience and by a General Surgery consultant with ten years of surgical experience. The operation was carried-out under general anesthesia without anesthetic complications. Laparotomy had confirmed some of the findings of the CT scan. Ileal and jejunal loops were dilated, ileal luminal stenosing, transitional, and ulcerating zone, and a mass measuring 3x3x2.5 cm distancing approximately 70-cm proximal to the ileocecal valve were found. Furthermore, several surrounding mesenteric lymph nodes were noted to be enlarged. However, there was no intussusception, rather the lobulated mass was pulling the mesentery inwards masquerading for intussusception. Excision the ileal loop which contained the mentioned zone along with the mass adherent to it was indicated in addition to a 3-cm free margin and a side-to-side/ileal-ileal anastomosis was done. All the specimens were sent for histopathological analysis.

Preliminary histopathology examination revealed presence of sheets and masses of small monomorphic lymphocytic cells showing mild atypia invading the superficial muscular wall. No evidence of lymph node involvement was seen. Surgical resection lines (Proximal and Distal) were free. Diagnosis is compatible with intestinal lymphoma (Fig. 2A-B-C-D).

Immunohistochemistry revealed a Diffuse Large B-Cell Lymphoma (DLBCL) involving all the layers of the ileal wall in addition to the presence of surrounding infiltrates. (CD20) positive. (Fig. 3A–B) and (CD3) negative (Fig. 3C–D).

The patient had an uneventful postoperative recovery and has been followed-up in the outpatient clinics for 10 months following his operation and was referred to a specialized oncology tertiary university hospital where he received adjuvant chemotherapy of (R-CHOP). He received a total of 8 regimens of R-CHOP, the last of which was 7 months postoperatively. He registered regular and scheduled visits with the General Surgery clinic and the oncology clinic to monitor his disease progression and evaluate his response to treatment. Evaluation included laboratory tests, physical examination, abdominal ultrasounds, an echocardiogram, and several postoperative CT scans of the Chest/Abdomen/Pelvis. Postoperative CT scans of the Chest/Abdomen/Pelvis revealed the following findings: Chest was clear and free of any reactive lymphadenopathy- No pleural effusion- Mediastinum was normal. Liver/Spleen/Pancreas/Kidneys/Bladder/Pelvis were normal. No presence of lymphadenopathy surrounding the abdominal aorta -as indicated by the arrows- (Fig. 4A-B-C-D-E).

3. Discussion

B-cell lymphoma is the third most frequent neoplasm of the intestine and makes-up merely 0.5% of all colonic neoplasms. [11] The close estimate of patients with such a neoplasm who require emergent surgery is 20% [12]. This neoplasm primarily involves old age groups of the population with a mean age being older than 55 years old and it has a higher tendency to affect males rather than females [13–14]. The most frequently involved site in the small intestine is the ileum followed by the jejunum, and the duodenum (6–8%) [14].

The most prevalent symptom in patients who present to the hospital is abdominal pain, followed by loss of appetite. This is true in both types of lymphoma; gastric and intestinal (ranging approximately 78% (gastric lymphoma) to 53.3% (when several segments of the gastrointestinal tract are involved). Obstipation and intestinal ileus are the next most common presenting symptoms in patients with intestinal lymphoma. Sometimes, fecal occult blood is positive in those patients, but most frequently, the gastrointestinal bleeding is macroscopic when the compromised segments are stomach or the ileocecal. Perforation is a rare presentation accounting for 1.8% in gastric lymphoma and 9.4% in NHL of the small bowel). Cardinal/B -symptoms happened in an approximate range of 11.9% for gastric lymphoma to 25% in multiple gastrointestinal sites. Weight loss was not categorized as a cardinal symptom. Nevertheless, it was considered as a direct result of the lymphoma [5].

B-cell lymphomas test positive for B-cell markers (CD19, CD20,

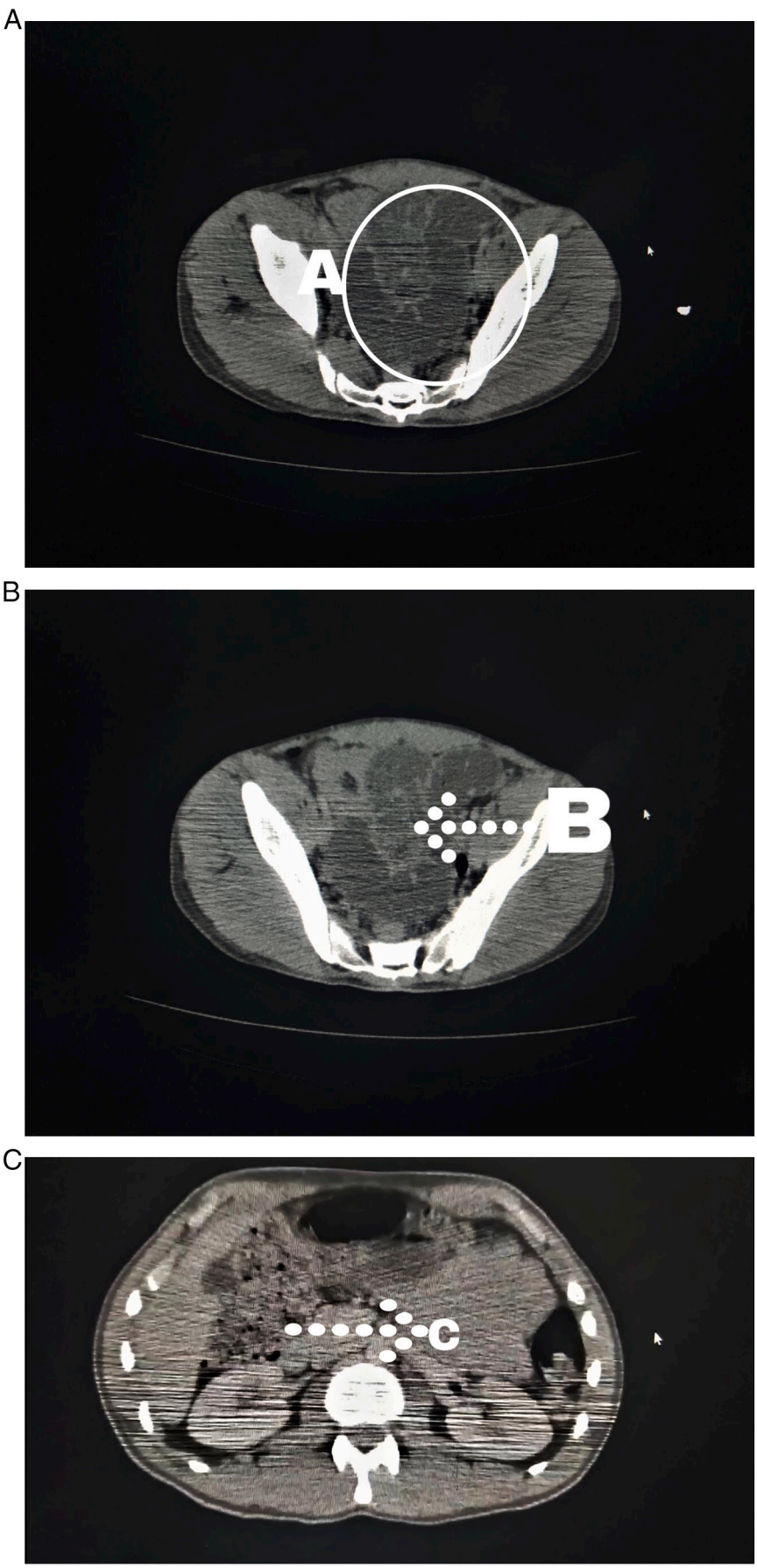

Fig. 1. A–B CT scan of the abdomen and pelvis showing thickening of the small bowel loops in the right iliac fossa. Moreover, there is ileal and jejunal loops dilation (Circle A), in addition to what appears to be an invagination of the small bowel mesentery (Arrow B)
C CT scan of the abdomen and pelvis showing A mass-like structure with lobulated borders situated anterio-inferior to the left kidney compressing the small bowel loops proximal to the ileocecal valve (Arrow C).

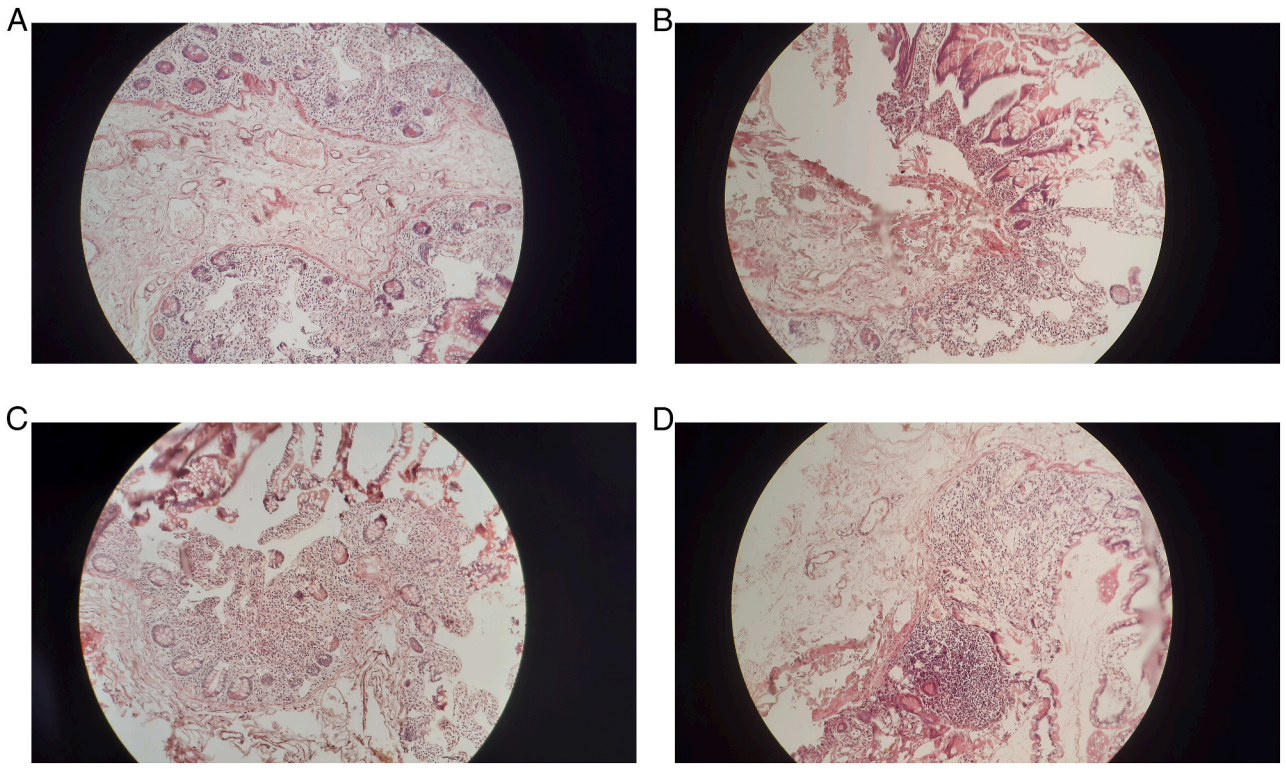

Fig. 2. A-B-C-D H&E stain showing tall and large villi at the surface, plus, focal ulceration with presence of sheets and masses of small monomorphic lymphocytic cells showing mild atypia invading the superficial muscular wall.

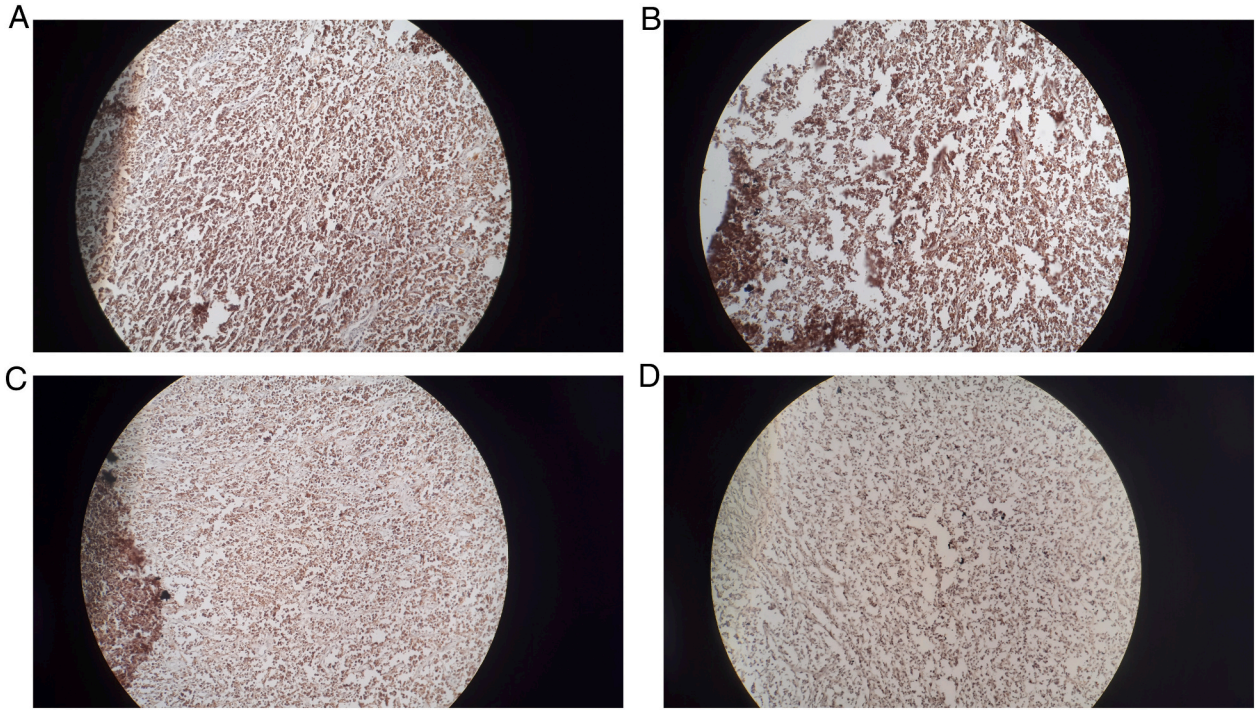

Fig. 3. A–B Immunohistochemistry stain revealed CD20 marker to be positive. C-D Immunohistochemistry stain revealed CD3 marker to be negative.

CD22, and CD79α). In contrast, T-Cell lymphomas yield positive for T-cell markers such as CD2, CD3 (considered lineage specific; B-cell or T-cell), CD5, CD7, CD4, and CD8 [15].

In comparison to the sole use of surgical treatment, adjuvant chemotherapy and/or radiotherapy can immensely raise the chances of survival. A Danish research group dedicated for lymphoma has established that surgical treatment in addition to adjuvant chemotherapy is more efficiently thorough than any distinct therapeutic modalities used

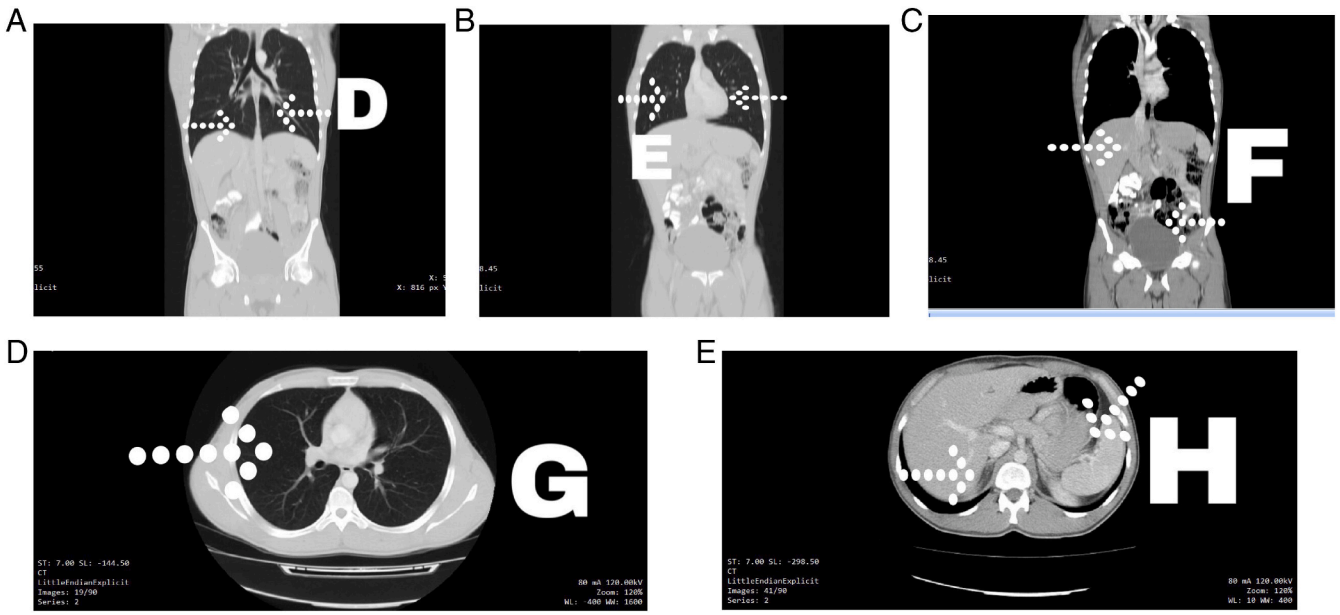

Fig. 4. A Post-Op CT scan/Coronal view of the Chest showing clear lung field (Arrow D)
B Post-Op CT scan/Coronal view of the Chest showing clear lung field (Arrow E)
C Post-Op CT scan/Coronal view of the Abdomen and Pelvis showing clear negative signs for tumor (Arrow F)
D Post-Op CT scan/cross sectional view of the Chest showing clear lung field (Arrow G)
E Post-Op CT scan/cross sectional view of the Abdomen and Pelvis with focus on the Liver and its hilum, Pancreas, and Spleen with its hilum, showing negative signs for tumor (Arrow H).

together with regards to localized neoplasm [16].

The mainstream and up-to-date chemotherapeutic management is (R-CHOP). DLBCL studies concluded that it can be cured in greater than 60% of patients who underwent treatment with the combination therapy R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) [17].

Unified management with surgical excision in addition to adjuvant chemotherapy showed to significantly raise the overall survival rates for small bowel large B-cell lymphoma [18] Radiotherapy is of no benefit when it comes to treatment of DLBCL involving the small intestine [19].

4. Conclusion

Patient presentation varies, and the symptoms are ill-defined. B-Symptoms combined with obstipation in an adolescent male should focus our attention to this type of malignancy. Nonetheless, these obscure and non-characteristic symptoms result in this type of tumors being clinically unrecognizable from other occurring gastrointestinal malignancies Additionally, we must be mindful that this neoplasm could present with the radiological findings consistent with another pathology. This may lead to utilizing different treatment modalities which would be ineffective because the disease had radiological findings that suggest an entirely different illness. Although rare, we should consider this diagnosis in an adolescent male presenting with a similar clinical picture. This will allow us to make better timely judgements and will help us perform therapeutic interventions swiftly to limit morbidity and mortality.

Abbreviations

DLBCL	Diffuse Large B-Cell Lymphoma
NHL	Non-Hodgkin's Lymphoma
CT	Computed Tomography
ED	Emergency Department
H&E	Hematoxylin and Eosin
R-CHOP	Rituximab- Cyclophosphamide- Hydroxydaunorubicin- Oncovin- Prednisone

Ethics approval

Institutional review board approval is not required for deidentified single case reports or histories based on institutional policies.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Availability of data and materials

The datasets generated during and/or analyzed during the current study are not publicly available because the Data were obtained from the hospital computer-based in-house system. Data are available from the corresponding author upon reasonable request.

Sources of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Research registration

N/A.

Guarantor

Reham Albrijawy.

CRediT authorship contribution statement

OA, RA: conceptualization, resources, who wrote, original drafted, edited, visualized, validated, and literature reviewed the manuscript.

JS, FA: supervision, project administration, and are the surgeons who performed the operation and reviewed the manuscript.

AA: specialist, General Surgery supervision, who supervised the operation.

RA: the corresponding author who submitted the paper for publication.

All authors read and approved the final manuscript.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

The authors declare that they have no competing interests.

Acknowledgements

-Lina Haffar's Histopathology and Immunohistochemistry Laboratory, Damascus, Syria. For their role in the histopathological analysis of the specimens.

-Radiology Department at Al-Mouwasat University Hospital, Damascus, Syria. For their role in radiological imaging and result interpretation.

-Dr. Amir Adi, MD, Department of Radiology at Al-Mouwasat University Hospital, Damascus, Syria. For his role in radiological interpretation of the CT scan results.

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