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A case of multiple recurrent intussusceptions due to multiple lymphomatous polyposis associated with diffuse large B-cell lymphoma of gastrointestinal tract in a 15-year-old child: A rare case report

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Multiple lymphomatous polyposis (MLP) is a distinctive and rare entity of primary gastrointestinal (GI) lymphoma characterized by polypoid lymphomatous tissue in long segments of the gut and a strong tendency for spread throughout the GI tract. Although many cases of MLP presenting as intussusceptions in adults have been reported, we report a rare case of multiple recurrent intussusceptions due to MLP associated with high-grade Diffuse Large B-cell lymphoma (DLBCL) of the entire GI tract in a 15-year-old child.

CASE PRESENTATION: A 15-year-old child previously operated for acute intestinal obstruction, presented with intermittent abdominal pain, nausea and vomiting. Imaging studies confirmed the diagnosis of multiple small bowel intussusceptions. Patient was treated by exploratory laparotomy and multiple resection anastomosis. Histopathology confirmed the diagnosis of MLP due to DLBCL. The patient received chemotherapy following surgery. So far, at 6 months of follow-up, Patient is doing well.

CLINICAL DISCUSSION: Malignant tumors of the small intestine are unusual, with non-specific clinical presentation. Although ultrasound (US), CT, FDG-PET/CT and endoscopic evaluation are essential modalities for the diagnosis of intestinal polyposis. Final diagnosis of MLP can only be confirmed after histopathological examination and immunohistochemistry studies. Surgical resection followed by appropriate chemotherapy is the treatment of choice.

CONCLUSIONS: MLP due to DLBCL has rarely been described in young patients under the age of 18 years. We should keep a high index of suspicion for malignant GI lymphoma in cases of intussusception, especially in older children.

1. Introduction

Gastrointestinal (GI) tract is the most common extra nodal site for lymphoma constituting for 5%–20% of all cases. Primary GI lymphoma is very rare, constituting only about 1%–4% of all GI malignancies. Lymphoma can arise from any part of the GI tract, Stomach (50–60%) is the commonest site followed by small intestine (20–30) while colon, rectum and oesophagus (<1%) account for a minority of cases [1]. Grossly, Primary gastrointestinal lymphomas are classified as solid tumor type which is most common, while diffuse infiltration type and polyposis type are rare. Histopathologically, approximately 90% of primary gastric lymphomas and 60%–80% of intestinal lymphomas are B-cell type followed by T-cell NHL and Hodgkin's lymphoma [2].

Multiple lymphomatous polyposis (MLP) is a rare and distinct form of gastrointestinal lymphoma with tumor cells forming polypoid tumors in long segments of the GI tract range from 0.2 to 2 cm in size [3]. The most common lymphoma subtype presenting as MLP is mantle cell lymphoma (MCL), a mature, aggressive B-cell lymphoma [4]. DLBCL is a heterogeneous entity rarely causing acute obstructive symptoms and intussusceptions [5].

Intussusception is a pathological condition in which telescoping of a proximal segment of bowel occurs into lumen of the distal segment. About 90% of cases of intussusception in children arise from unknown causes, which include infections, anatomical factors and altered motility. Only 10% of them are caused by a pathological lesion (for examples - Meckel's diverticulum, polyp, and benign or

Abbreviations: MLP, Multiple Lymphomatous Polyposis; DLBCL, Diffuse Large B-cell lymphoma; CT, Computerized tomography; FDG-PET/CT, Fluorodeoxyglucose -Positron emission tomography.

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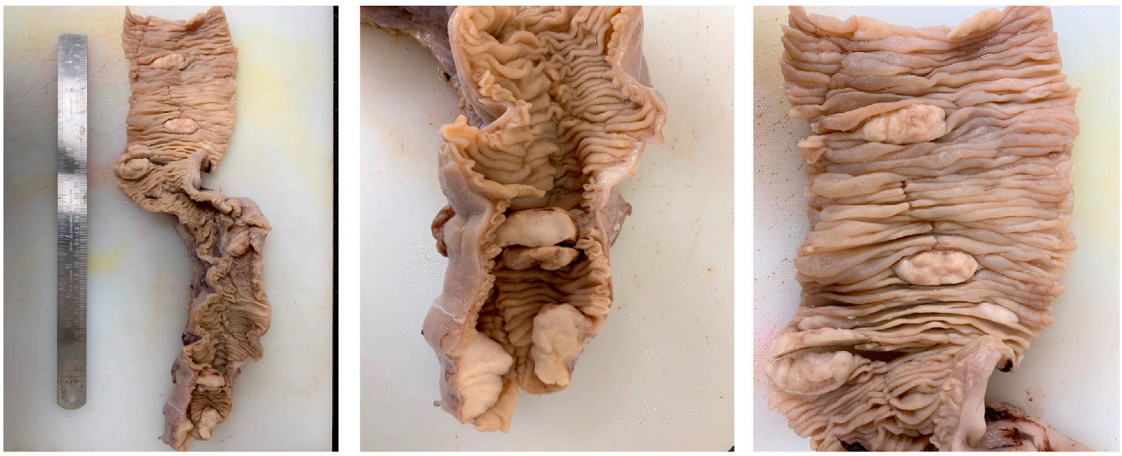

Fig. 1. Perioperative photographs showing Fig. 1 (a) more than 45 cms long resescted specimen showing multiple sessile polyps (leading point) with sites of multiple intussusception. Fig. 1 (b) Magnified image of a segment of small bowel showing multiple studded polyps Fig. 1 (c) Multiple Lymphomatous Polyposis -sessile polyps -largest approximately 4–5 cm size with congested mucosa and patchy gangrenous changes.

malignant tumor [6]. Majority of lead points in the small intestine consist of benign lesions with malignant lesions accounting for up to 30% of cases of intussusception [7]. We herein present a rare case of multiple jejuno-jejunal and ileo-ileal intussusceptions with MLP caused by DLBCL in a 15-year-old child who underwent surgical intervention followed by chemotherapy. The kind of presentation as MLP by DLBCL causing acute intestinal obstruction due multiple intussusceptions has never been described.

2. Material and methods

In this paper, we present and discuss a rare case of multiple intussusceptions caused by multiple lymphomatous polyposis with its successful management. In addition, for this review, a search of the English medical language literature in Pubmed and Google Scholar was conducted for every case report, series, letter to the editor, original article, and literature review related to multiple lymphomatous polyposis. Key words used were multiple lymphomatous polyposis, intussusception and lymphoma. A Surgical team consisting of first two authors and lead by corresponding author performed the procedure in a district corporate hospital. The experience of the lead surgeon is more than 15 years in the gastrointestinal and hepatobiliary department. This case report has been reported in line with SCARE Criteria [8].

3. Case report

A 15-year-old boy admitted in a district hospital with 2 weeks history of intermittent abdominal pain and nausea with vomiting, initially diagnosed with sub-acute intestinal obstruction and was operated on 20 February 2020. Intraoperatively multiple polyps in small intestine and stomach with multiple intussusceptions were noted, however histopathology examination report was not performed. Subsequently, His symptoms progressively worsened to severe anorexia, weight loss, constipation, and bilious vomiting due to recurrent intestinal obstruction. The patient was then referred to our hospital in a private ambulance in May 2020. The patient had no prior history of allergy, lymphoproliferative disorder or taking any immunosuppressive drugs. Family history was also insignificant in terms of inheritable conditions. Physical examination revealed midline laparotomy scar with abdominal tenderness in the right lower abdomen. Patient had hypoproteinemia (serum albumin 1.7 g/dl), anaemia (haemoglobin level of 77 g/dl) with a haematocrit of 29% and white blood cell count of 13.5 × 109 cells/litre (63% neutrophils). USG showed multi segmental intussusceptions with mesenteric lymphadenopathy. A contrast computed tomography (CT) scan of abdomen revealed multiple heterogeneously enhancing nodular mucosal thickening in stomach, small bowel loops and ascending colon till hepatic flexure, bilateral pleural effusion and ascites.

His COVID-19 RT-PCR test report was negative. The patient was treated with blood transfusion and intravenous albumin prior to the intervention. Haemoglobin stabilised at 10.1 g/dl after that, he was taken for exploratory laparotomy. Intra-operatively five Jejuno-jejunal and ileo-ileal intussuceptions was noted over multiple sessile polyps (leading point) -largest approximately 4–5 cm in size with congested mucosa and patchy gangrenous changes [Fig. 1a–c]. The whole small bowel and stomach were studded with multiple sessile polyps. We performed three segmental resection and end-to-end anastomosis. Two abdominal drains were placed in pelvis and subhepatic region. Patient was extubated in operating room and kept in intensive care unit overnight. Patient received intravenous antibiotics, fluids, analgesics and 20% albumin during post-operative course. Patient postoperative course was uneventful and tolerated the procedure well. He was discharged on 10th postoperative day.

The gross histopathological examination of the polypoid lesion showed features of diffuse large B-cell Non Hodgkin lymphoma (DLBCL, Activated B-like phenotype) with following immunohistochemistry markers CD20 (B-cell marker): Strong & diffuse membranous staining, MIB-1 labelling index: 70–75%, CD3 (T-cell marker):Negative (Positive in background small lymphocytes), Bcl6 (Germinal centre marker): Negative, CD-10 (Germinal centre marker): Negative, MUM-1/IRF (post-germinal centre or activated B-like): Positive, Pan-Cytokeratin & CD-30: Negative [Fig. 2a–d].

Patient was then referred to medical oncology for postoperative chemotherapy (Rituximab-CHOP). The child is doing well so far after 6 months of surgery with regular monthly follow-up. Outcomes were measured by history, examination, relevant blood investigations and ultrasound of abdomen. Now he is under the follow-up of medical oncology department.

4. Discussion

Primary malignant tumors of the small intestine account for less than 2% of all gastrointestinal malignancies [9]. Lymphoma

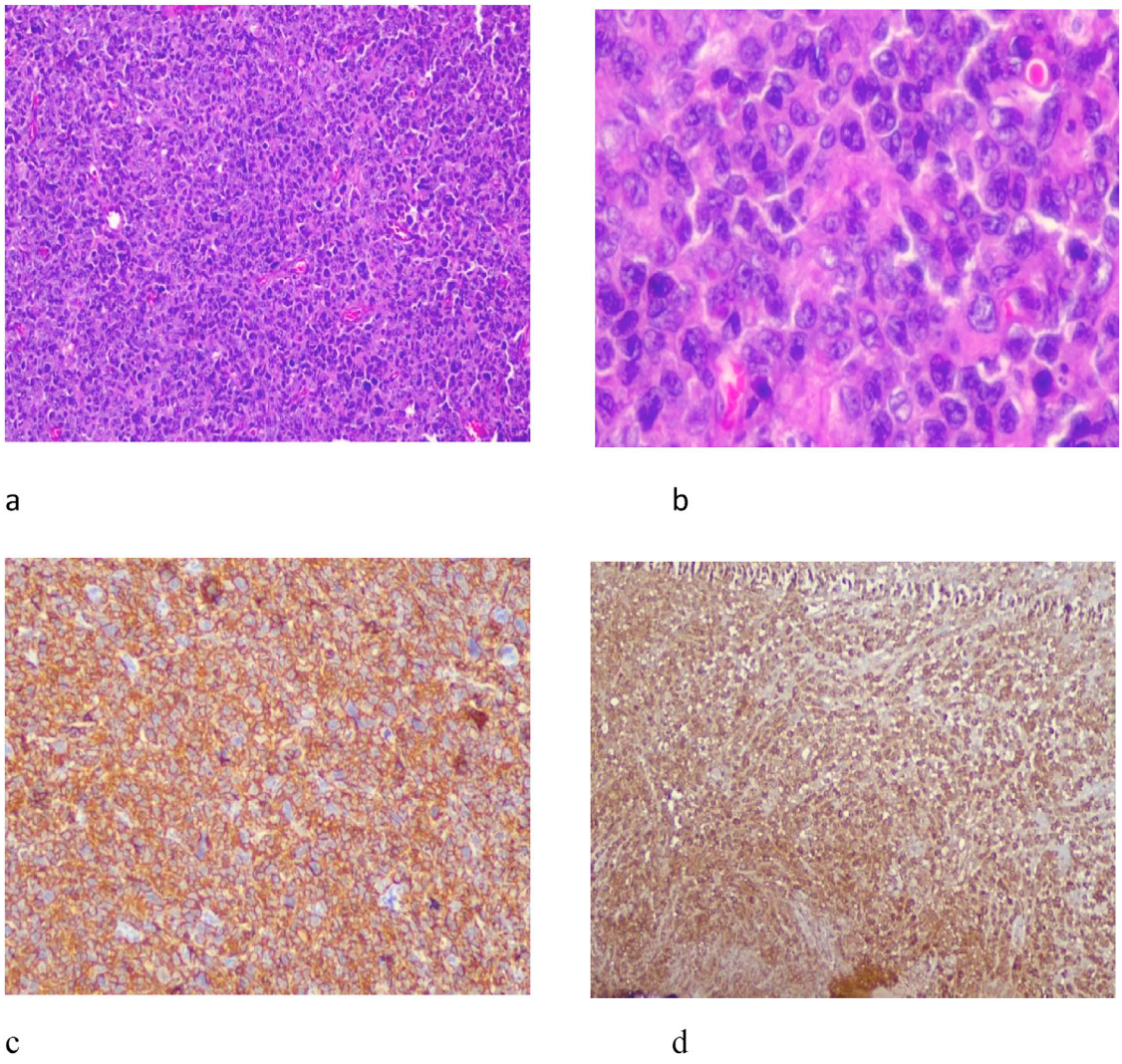

Fig. 2. Section examined show a malignant tumor composed of diffusely infiltration by large atypical cells admixed with small mature lymphocytes and reactive histiocytes, tumor cells exhibit basophilic cytoplasm round/indented/irregularly folded nuclei vesicular chromatin and prominent nucleoli. Mitotic figures are readily found and evidence of prominent karyorrhexis. Diffuse large B cell lymphoma; Activated B cell phenotype (ABC-DLBCL). Fig. 2a: showing B-cell lymphoma cells in the intestinal mucosa (×10 H & E stain). Fig. 2b: Diffuse large B cell lymphoma 20× H& E stain. Fig. 2c: Strong cell membrane positive staining reaction for CD-20 (10×). Fig. 2d: Positive KI 67 (MIB-1 labelling index: 70–75%).

constitutes 15%–20% of all small intestine neoplasms. Ileum is far the most common site (60%–65%) followed by jejunum (20%–25%), duodenum (6%–8%) and other sites (8%–9%) [1,10].

Ever since MLP was first described by Cornes in 1961, It has not been explored in a large series of patients until, A French study group conducted a Prospective Clinicopathologic Study of 31 Cases of MLP among the 350 cases of primary gastrointestinal lymphoma. Their mean age was 55 years (range, 21–79 years) with male/female ratio of 2.6:1 [11]. MCL is the most common lymphoma subtype that presents as MLP. Successive studies showed that follicular lymphoma, MALT lymphoma, DLBCL and small lymphocytic lymphoma may also be present as MLP [4,12].

Another study reviewed thirty-six published cases of intussusceptions caused by lymphoma. Patients were aged from 16 to 86 years. Only 7 patients out of 36 from age group of 16–24 presented with Intussusceptions secondary Non-Hodgkin lymphoma [13].

Our patient is distinctive in that the DLBCL with MLP manifested as an unusual presentation of recurrent intussusceptions with infrequently large polyps (~5 cm) in a paediatric patient.

The clinical presentation of small intestinal lymphoma is non-specific symptoms, such as colicky abdominal pain, nausea, vomiting and weight loss. Seldom there are acute obstructive symptoms, intussusceptions, perforation or diarrhoea [14]. The age of presentation varies with the histological subtype of lymphoma.

Diagnostic modalities for evaluation of the small intestinal polyps or tumors are ultrasound (US), CT, magnetic resonance imaging (MRI), enteroclysis, endoscopic procedures (EUS), diagnostic laparoscopy, scintigraphy, angiography and FDG-PET-CT. The classical imaging characteristics of Intussusceptions include the target or doughnut sign in the transverse view and the pseudokidney, sandwich, or hayfork sign in the longitudinal view. Abdominal Contrast-enhanced (CE) CT is currently considered the most sensitive radiological technique for confirming intussusceptions. CECT scan of the chest, abdomen and pelvis is employed to stage gastrointestinal lymphomas with a marked sensitivity and specificity. Capsule endoscopy (CE) is a non-invasive diagnostic test used in patients without obstruction to locate the source of gastrointestinal bleeding and identify the cause of other intestinal disorders, including intussusception and various tumors. Small intestine lymphoma appears as a mass, polyp and ulcer on CE [1,15]. FDG-PET has a significant advantage in staging of DLBCL, follicular lymphoma and MCL with a sensitivity of 80% and a specificity of 90% [16].

Table 1
International prognostic index.

Adverse risk factors
- Age > 60 yr
- ≥ 2 extranodal sites
- Ann arbor stage III-IV
- Performance status ≥ 2 (ECOG)
- High lactate dehydrogenase

Risk
- Low risk (0–1 points) - 5-year survival of 73%
- Low-intermediate risk (2 points) - 5-year survival of 51%
- High-intermediate risk (3 points) - 5-year survival of 43%
- High risk (4–5 points) - 5-year survival of 26%

A study conducted by Seok Jin Kim et al. concluded [17] that patients with localized disease, surgery plus chemotherapy (Rituximab with cyclophosphamide, doxorubicin, vincristine, and prednisolone R-CHOP) yielded a lower relapse rate than did chemo therapy alone. Surgical resection followed by chemotherapy might be an effective treatment strategy with acceptable quality of life (QOL) deterioration for localized intestinal DLBCL. In general, chemotherapy is recommended along with surgery in cases with poor prognostic factors, such as age of > 60 years, ECOG performance status of ≥ 2, high LDH level-cell phenotype, extra nodal involvement of ≥ 2 and Ann Arbor stage of III to IV. Higher positivity rates of the Ki67 proliferation index immunochemically associated with the aggressive course of a tumor (Table 1). However, the extent of the surgical procedure should be determined according to intra-operative findings in cases requiring urgent surgery because of signs and symptoms of intestinal obstruction.

The prognosis of MLP is poor due to its accelerated proliferation. Most of the patients had advanced diseases at the time of diagnosis. Long-term follow-up is necessary to predict the outcome [18].

5. Conclusion

Primary GI lymphoma should always be kept in the differential diagnosis in cases of Intussusception, especially in the older age group of children. The most appropriate approach is to perform surgical resection and immunohistochemistry for localized intestinal DLBCL. Adjuvant chemotherapy is an effective treatment strategy in such cases.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

Ethical approval not required.

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.

Authors' contribution

Kapil Dev Sharma helped in analysis and interpretation of data, revising the article critically for important intellectual content and final approval of the version to be submitted. Ashish V. Massey helped in conception and design of study, analysis and interpretation of data, drafting the article and final approval of the version to be submitted. Manish Vijayvargiya helped in acquisition of data, revising the article critically for important intellectual content and final approval of the version to be submitted. Sundeep Jain helped writing, editing, analysis and interpretation of data, revising the article critically for important intellectual content and final approval of the version to be submitted.

Registration of research studies

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CRediT authorship contribution statement

Kapil Dev Sharma: Data curation, Investigation, Project administration. **Ashish V. Massey:** Conceptualization, Methodology, Software, Data curation, Writing - original draft. **Manish Vijayvargiya:** Investigation, Validation. **Sundeep Jain:** Writing - review & editing, Visualization, Supervision.

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