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Case Report

Adult Burkitt's lymphoma presenting as ileocecal intussusception

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ABSTRACT

Most lymphomas of gastrointestinal tract arise mainly in stomach but can affect small bowel. Burkitt's lymphoma stands for 9% of the cases of primary small intestine lymphoma and clinical presentations may vary. Case report of a 58-years old man presenting an acute abdomen secondary to intestinal obstruction with tomography revealing an ileocecal mass suspicious for intussusception. Surgery was performed and an invaginating mass from ileum-to-cecum lead to a laparoscopic right colectomy. Histopathological analysis established the diagnosis of ileocecal Burkitt's lymphoma. Post-operative period was uneventful and the patient was referred to oncohematology and started early chemotherapy. Clinical situations of intestinal intussusception or small-bowel masses presenting with obstruction should rise the suspicion for malignancy and the surgeon must be aware of this clinical entity to proceed with an oncologically safe surgery. Intestinal perforation may indicates surgery as it leads to peritoneal tumor spread and indicates a worse prognosis. Laparoscopic surgery approaches must be seriously considered in selected cases. In intestinal lymphoma, early refer to oncohematology for evaluation and treatment is essential and establishes a better prognosis.

Keywords: Intussusception, Burkitt, Lymphoma, Small intestine, Case report, Ileum, Cecum

INTRODUCTION

Most malignant lymphomas of gastrointestinal tract arise mainly in stomach, with reports indicating that over 90% are B-cell lymphomas and mucosa-associated lymphoid tissue lymphoma.[1] Burkitt's lymphoma (BL) is a B-cell lymphoma, a form of non-Hodgkin's lymphoma and the incidence in the small intestine is low. It is characterized by aggressive growth and rapid development of clinical features.[2]

BL stands for 9% of the cases of primary small intestine lymphoma and clinical presentations includes intestinal obstruction or intussusception, small bowel perforation or as an incidental finding.[1-7] We presented a case of intestinal obstruction by the form of ileum to cecum intussusception secondary to a Burkitt's lymphoma in a middle-aged man with signs of microperforation.

CASE REPORT

A 58-years old man was admitted to our hospital's emergency department with symptoms of nausea, vomiting, recurrent abdominal pain and anorexia of 10 kg in 5 weeks. Chronic diseases includes arterial hypertension, diabetes mellitus type 2 and chronic obstructive pulmonary disease. Physical examination revealed blood pressure 132/87 mmHg, temperature 37.3 °C, and a pulse rate of 82 bpm. He had localized tenderness in lower right abdomen where a mass was palpable. No further alterations in physical exam. Blood count with normal levels of leukocytes, hemoglobin and

platelet count. C-reactive protein was elevated 5X the superior limit. There was no evidence of organ dysfunction. He performed an abdominopelvic computed tomography that revealed an intussuscepted ileal mass to the cecum with suspicion for microperforation, parietal thickening of ascending colon wall and ileum distention. He was proposed for urgent surgery and we chose a laparoscopic approach. We used an umbilical port of 11 mm for celioscopy and work ports in left flank, lower left and lower right abdomen. An invaginating 80 mm mass from ileum-to-cecum was found and a laparoscopic right colectomy with extracorporeal primary anastomosis was performed (Figures 1 and 2). No peritoneal soiling was observed. No perforation was observed in free bowel or colon. Subsequent removal of the specimen was performed via mini-Pfannenstiel incision that was used to work on the anastomosis (Figures 2 to 4). An abdominal wall protector was used for extracorporeal handling thus preventing surgical site infection and tumor seeding.

The patient recovered well and was discharged on post-operative day 5. Oral intake, bowel transit and pain control was accomplished during this time. No surgical complications were registered. The histopathological analysis resulted in B-cell lymphoid proliferation with immunochemistry positive for CD20, CD-79-alpha, CD10, Bcl-6 and with a Ki-67 near 100% (Figure 5 and 7). The 'starry-sky' appearance, a land-mark in histologic diagnosis was also present (Figure 6). Microperforation was macroscopically observed inside of invaginated segment. Therefore, we established the diagnosis of ileal BL and referred the patient to oncohematology for early evaluation and treatment.

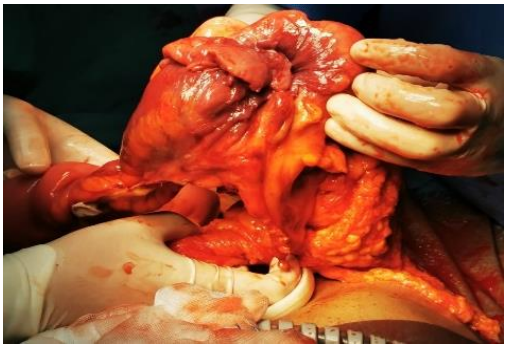

Figure 1: Ileocecal mass before extracorporeal transection.

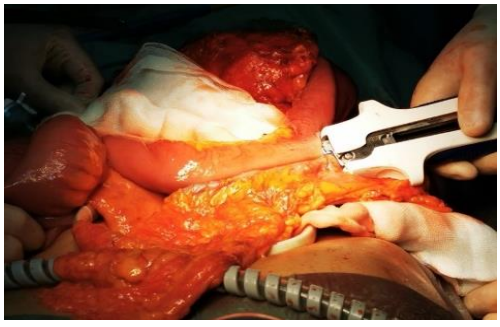

Figure 2: Extracorporeal anastomosis.

Figure 3: Surgical specimen-inferior view.

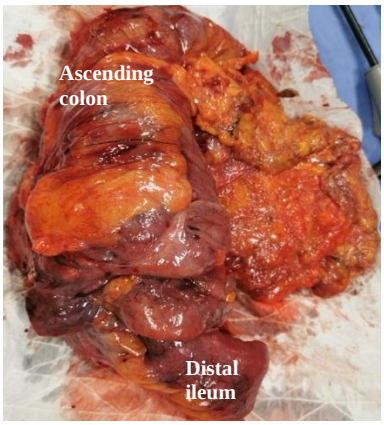

Figure 4: Surgical specimen-anterior view.

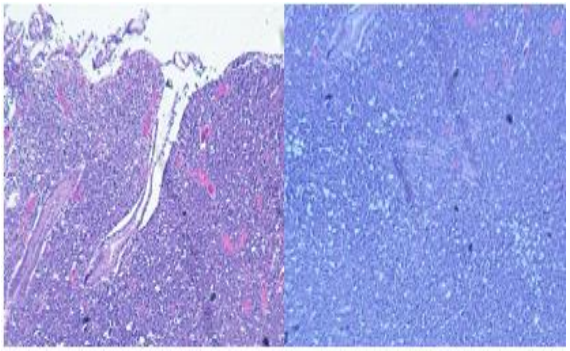

Figure 5: Histology: lymphoma-mucosal destruction (40X magnification).

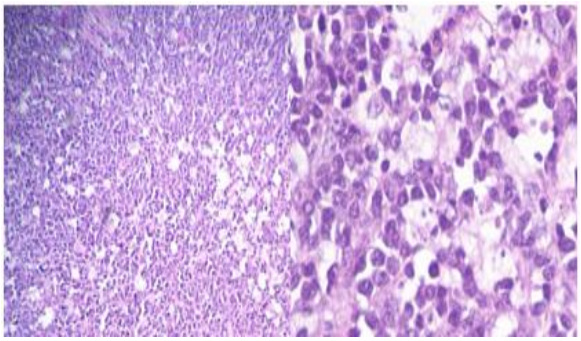

Figure 6: Histology: the classic 'starry sky' appearance caused by macrophages-intracitoplasmatic debris (100X and 400X magnification).

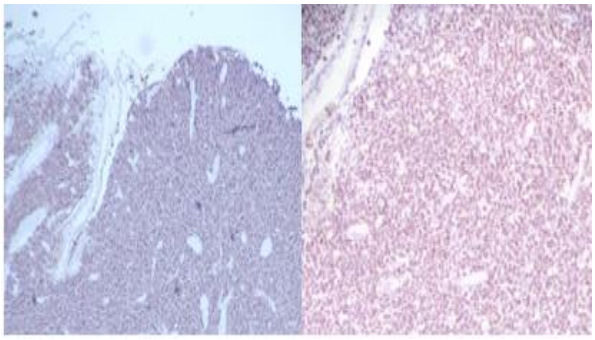

Figure 7: Immunochemistry + for CD20 (left) and Ki-67 nuclear marking near 100% (right).

A lumbar puncture was performed and revealed central nervous system infiltration and massive medullar invasion. The patient was treated with R-HyperCVAD and toxicity was observed: severe infectious complications secondary to neutropenia and spontaneous subdural bleed with 5 hospital admissions in the first year of treatment. Currently in late second year of follow-up, a medullar, leptomeningeal and imaging complete remission was accomplished. The patient also maintains absence of symptoms, normal blood analysis and a good and sustained quality of life.

DISCUSSION

Intestinal lymphoma is rare and BL is also a rarity in this spectre. Primary presentation as an intussuscepted microperforated mass from ileum to the cecum is scarce in reported literature.

Panaccio et al reported a case of intestinal obstruction due to BL causing ileocecal invagination treated with laparoscopic right colectomy with extracorporeal anastomosis, as in our report. Although, no microperforation was reported, unlike our case.[3]

Small bowell perforation due to BL literature is scarce, with the first reported case registered in 2021 by Takayama et al.[2] As pointed by the authors, there are several articles reporting gastrointestinal perforation occurred during chemotherapy in BL, but the first case reporting intestinal perforation due to BL was by Takayama's team. A midline laparotomy approach was used. The role of surgery in intestinal BL remains controversial as it has a good response to chemotherapy by inducing rapid tumor regression and offers long-term remission.[8] Nevertheless, Magrath et al suggested that an aggressive operative debulking treatment before chemotherapy may increase survival.[9] In our situation, we had an invaginated mass with microperforation, which dictates surgery by intestinal obstruction status and two additional points related to peritoneal soiling - risk of peritonitis and risk of tumor seeding. It has been reported that cases of BL associated with perforation have a poorer prognosis.[10] Knowing that, surgery is mandatory in order to rescue the patient from acute setting of obstruction and perforation and to avoid peritoneal spreading. In this matter, our option for laparoscopic approach resulted in a favorable outcome. In terms of surgical approaches, there are no evidence leading to which surgical technique (laparoscopic or laparotomic) is better. This choice should be made based on clinical judgment and surgical team's experience.[3] Laparoscopic approach tends to minimize post-operative complication rates and costs, promotes an early hospital discharge and must always be taken as an option if surgical safety is guaranteed and the surgical team has laparoscopic expertise, as we know from population-based study of outcomes in emergent laparoscopic colectomy reported by Keller et al establishing that consideration for a laparoscopic approach in non-elective settings should increase.[11]

As supported by Takayama et al a prompt involvement of oncohematology after the diagnosis to initiate chemotherapy as early as possible after surgery, are thought to improve prognosis.[2] In our opinion, laparoscopic approach also has a role endorsing a better prognosis as it promotes faster recovery from surgery compared to open approach.

CONCLUSION

Small-bowel obstructing masses presenting as intussusception should rise the suspicion for malignancy and the surgeon must be aware of differential diagnosis, how to treat each case and ensure a prompt refer to oncologic medical team. Laparoscopic approach results in reduced post-operative complications and length of stay and must always be considered if an oncologically safe surgery may be performed respecting vascular and oncologic principles in the setting of a suspicious tumor with need for emergent treatment. Knowing that early as possible chemotherapy is an important prognostic factor in intestinal lymphoma, in our understanding laparoscopic approach should also has a role as a recovery booster in comparison to open approach and consequently promoting a better prognosis, without compromising surgical oncologic principles. Moreover, the presence of microperforation indicates a surgical approach with tumor resection as peritoneal soiling may lead to a worse prognosis.

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