

Case report

Non-Hodgkin's lymphoma revealed by an ilio-colic intussusception in a Moroccan patient: a case report

Karim Ibn Majdoub Hassani^{1,&}, Hicham El bouhaddouti², Abdelmalek Ousadden², Amal Ankouz², Meryem Boubou³, Siham Tizniti³, Khalid Mazaz¹, Khalid Ait Taleb²

¹Department of general surgery (B), University hospital, Hassan II, Fes, Morocco; ²Department of general surgery (A), University hospital Hassan II, Fes, Morocco; ³Department of Radiology, Universitet hospital Hassan II, Fes, Morocco

[&]Corresponding author: Ibn Majdoub Hassani Karim, 16, Rue Dominique, AV abi horayra, Zohor I, 30000, Fez, Morocco. Tel: 00212665600722

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Abstract

Intussusceptions are rare but well-known causes of the small bowel obstruction in adults and an underlying cause is present in the majority of cases. Lymphoma's involvement of the ileum is one of the rare causes of intussusception. CT is a sensitive examination that diagnoses intussusceptions and provides an excellent pre-operative evaluation including possible extension and dissemination especially in intestinal lymphomas. The treatment is almost always surgical and the pathological study is needed for diagnostic confirmation. Authors present an unusual case of intestinal intussusception due to lymphoma of the terminal part of the ileum in a 49-year-old man. Computed tomography confirmed the diagnosis of intussusception and non-Hodgkin's lymphoma of B-cell was diagnosed by histological examination after surgical treatment. Primary intestinal lymphomas differ from gastric lymphomas in clinical features, treatment, and prognosis. They are not well characterized and the standardized concepts for their clinical diagnosis and management are absent. The aim of this rare observation is to shed light on NHL of the small bowel, its clinical and radiological diagnosis and its treatment especially in forms revealed by intussusceptions in adults.

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Background

An Intussusception is defined as the telescoping of one segment of the gastrointestinal tract into an adjacent one. It is a relatively a common entity in childhood in which it is presented as an acute illness. Most cases in childhood occur idiopathically. In adults, an underlying cause is present in 80% of cases. These causes include malignant process, lipomas and polyps as well as oedema and fibrosis from recent or previous surgery. Authors present an unusual case of intestinal intussusception due to a primary non-Hodgkin lymphoma (NHL) of B-cell of the terminal part of ileum. The primary ileal localization of NHL is rare, it occurs 23 to 30% of gastrointestinal lymphomas and approximately 5% of peripheral NHL [1]. It differs from gastric lymphomas in clinical features, treatment, and prognosis. Although substantial progress has been achieved in the diagnosis and treatment of gastric lymphomas in recent years [2, 3] primary intestinal lymphomas are not well characterized, and standardized concepts for their clinical diagnosis and management are absent. The aim of this observation is to shed light on NHL of the small bowel, its clinical and radiological diagnosis and its treatment especially in forms revealed by intussusceptions in adults.

Patient and case report

A forty-nine year old Moroccan patient with a medical history of repeated abdominal cramping pain especially in the right iliac fossa and a constipation of 1 year's duration, was presented to the emergency department because of a three days history of incomplete intestinal obstruction with an acute abdominal pain cramp-like localized in the right iliac fossa with no particular radiation and no aggravating factor. He had no fever, no vomiting and no bleeding. The patient was not using any specific medication. His medical history did not suggest any major disease, as well. He had no prior history of abdominal surgery or trauma.

The physical examination revealed a conscious man whose temperature was 37°C, a pulse rate 90 beat per minute (bpm), a blood pressure 120/70 mm Hg. The abdominal examination revealed tenderness with a large not reducible mobile mass measuring approximately 12cm in the right iliac fossa. There was no liver enlargement and no splenomegaly or lymphadenopathy. A rectal examination showed no stool. Vital signs and the remaining of the physical examination were normal. Initial management of the patient involved intravenous fluid resuscitation, nasogastric tube insertion, routine bloods, supine chest and abdominal x-rays.

Laboratory data revealed a hematocrit of 31%, hemoglobin of 10,6g/dl, white blood cells of 10100 elements/mm³, a blood urea of 0, 25 g/L, and a creatinine level of 10 mg/L. Coagulation, lipase, and Liver enzymes laboratory data were normal. A chest radiograph showed clear lung fields. A supine abdominal x-ray revealed multiple dilated small-bowel loops with air-fluid levels, but with no free air. Ultrasonography (US) of the abdomen revealed a typical image of an intussusception in the right iliac fossa with dilated small bowel loops and no free intra-abdominal fluid.

Abdominal computed tomography (CT) confirmed results of US and concluded to an ileocaecal intussusception (Figure 1). Based on the imaging findings, we made a decision to perform a laparotomy. During the surgical exploration, the peritoneal cavity was filled with 200cc of blood-stained serous fluid while numerous dilated loops of small bowel were present. There was an ileo-colic intussusception of 14 cm in length realizing a large mass of the right iliac fossa. There was no mesenteric adenopathies.

An ileo-coecal enbloc resection was performed with continuity restored by ileo-colic end-to-end anastomosis (Figure 2). The specimen examination revealed a tumor of the terminal part of ileum with no mesenteric adenopathies. No complications occurred and the patient was discharged on the fifth day postoperatively.

Figure 1: CT with Intravenous contrast material, showed a round mass lesion of soft tissue with fatty tissue within it in the right iliac fossa. This soft tissue strands within the fat represent mesenteric blood vessels within the invaginated mesenteric fat. Some dilatation of the small bowel is also seen

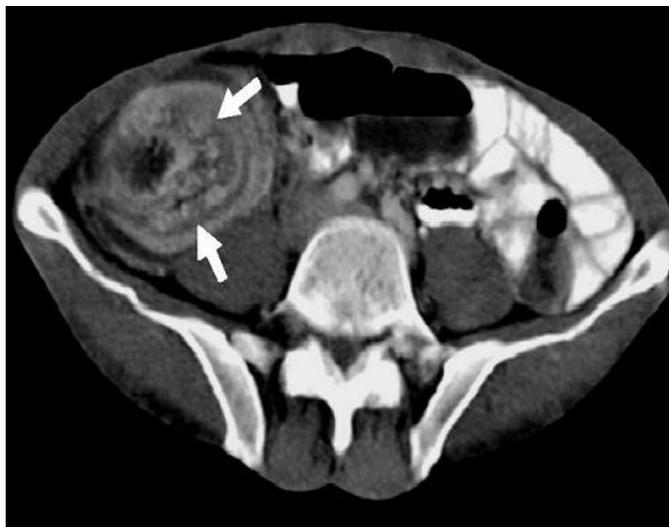


Figure 2: Specimen of the ileo-cecal resection



Histological examination of the specimen demonstrated non-Hodgkin's lymphoma of the diffuse large B-cell type (B-Cell NHL) which has a uniform, round-to-oval nuclei with vesicular chromatin and one or multiple conspicuous nucleoli (Figure 3).

Cells were positive for B-cell marker (CD20 +) at the immunohistochemical study (Figure 4). The disease was staged as primary ileal Stage 1 disease according to the Ann-Arbor classification. A postoperative chemotherapy (CHOP-R) stands for cyclophosphamide, hydroxydaunorubicin (Adriamycin), Oncovin (vincristine), and prednisolone combined with the monoclonal antibody rituximab was recommended but refused by the patient.

Figure 3: Histological examination demonstrating non-Hodgkin's lymphoma of the diffuse large B-cell type, which has uniform, round-to-oval nuclei with vesicular chromatin and one or multiple conspicuous nucleoli ($\times 200$).

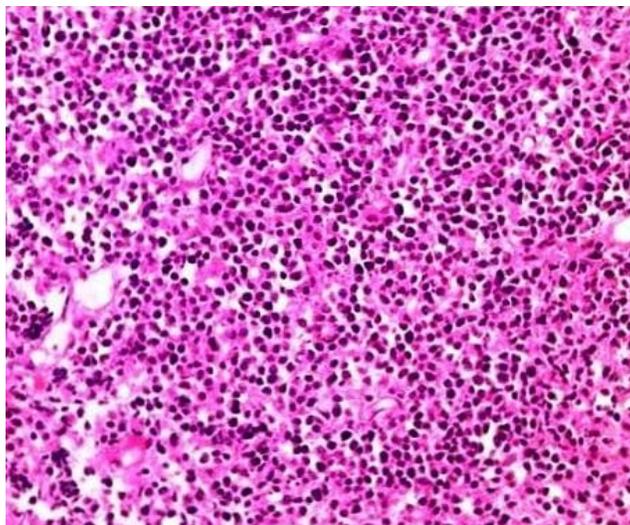
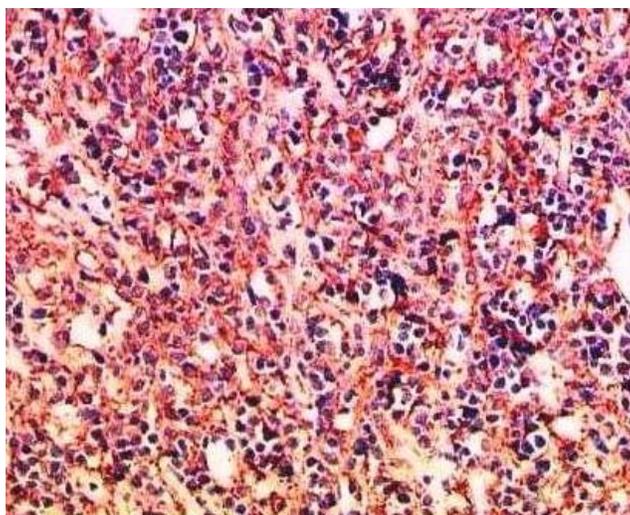


Figure 4: Photomicrograph showing cells positive for B-cell marker (CD20+) at the immunohistochemical study ($\times 200$)



Discussion

Intussusception is a rare disease in adults when compared with children; one case of adult intussusception for every 20 childhood ones [4]. In infants, intussusceptions are primitive in the majority of cases [5]; however, in adults, an organic lesion is found in 80% of cases mostly in the benign and malignant disease in the ileum and the colon. These organic lesions are represented by the Gastro intestinal stromal tumors (GIST), lipomas, polyps or adenopathies especially in the ileocaecal localization.

More rare are intussusceptions caused by lymphoma's involvement of the ileum. Few cases were reported in the literature [6]. Small bowel lymphomas are rare due to several reasons [7]; however, the incidence of this disease has been rising in recent years particularly among immunocompromised patients [8, 9]. Associations of small bowel B-cell lymphomas with post-transplantation, inflammatory bowel disease and some immunodeficiency syndromes have been reported [10, 11]. Nevertheless, one of the particularities of our case is that the medical history of our patient did not suggest any one of these diseases or syndromes. Intestinal T-cell non Hodgkin's lymphomas (T-Cell NHL) have been described as often multifocal and most frequently localized in the jejunum or proximal ileum. Many cases of association of intestinal T-Cell NHL to the celiac disease have been reported [1]. The primary follicular lymphoma of the small intestine is a distinct entity that originates from local antigen-responsive B cells whereas little is known about the pathogenesis of primary small bowel lymphoma.

A wide range of clinical presentations can occur for primary small bowel non-Hodgkin's lymphoma [12] including non-specific abdominal pain, ileus and weight loss. Diarrhea and fever are present in approximately one third of patients with T-Cell NHL but are rare in patients with B-Cell NHL [1]. Abdominal palpable mass is found in only 15% of cases [13]. Acute surgical conditions, such as perforation, bleeding and obstruction especially intussusception -which is the case of our patient- are initial complications which can be the revealing mode of the small bowel lymphomas. These initial complications occurred up to 70% of cases in recent prospective studies; the predominant complications in patients with B-Cell NHL and T-Cell NHL were intestinal obstruction and perforation, respectively [1].

Concerning the intestinal intussusceptions, it is defined by the telescoping and penetration of an intestinal segment in the downstream segment. Its evolutionary mode is usually sub-acute or chronic. Anatomically, in adults and regardless of the cause, the ileum is regarded as a preferential area of intussusceptions. The colo-colic intussusceptions present only 27% of cases and the colorectal forms are more rare [14]. The hyperperistaltism result from the presence of a pedunculated or non-pedunculated mass acted on an intestinal segment. Hyperperistaltism would be triggered by neurovegetatif reflexes and would be responsible for the formation of the width of intussusception which is the anatomicofunctional condition for the establishment of intussusceptions [15, 16].

Radiological diagnosis of intussusception especially the ileo-cecal one caused by lymphoma can be suspected in ultra sonography (US) which can show a typical image of intussusception, but the computed tomography (CT) appearance of it is characteristic. It helps diagnose obstructive syndrome, its mechanism, the presence of the intussusception, its precise location and show its causes. It can also detect the organic cause in 71% of cases [17]. The most common finding in CT is a thickened segment of bowel with an eccentrically placed crescent-like fatty area, representing the intussusception and the intussuscepted mesentery. They appear either as a round target mass or as an oblong sausage shaped mass. Another common finding is a rim-shaped accumulation of contrast material in the periphery of the mass. In addition, air bubbles in the uppermost part of the intussusception can be observed in some cases [18]. The role of CT is more important in cases of suspected abdominal lymphoma and polyps. It can objective a thickening of the wall associated with digestive adenopathies in lymphoma or a tissue density in the event of polyp.

Appropriate management of primary small bowel lymphoma is, therefore, still under discussion. One aspect of this discussion is that the surgical approach is necessary. Another aspect is that surgery is necessary but in combination with chemotherapy. According to few cases and to the absence of randomized trials in primary small bowel lymphoma, the optimal treatment strategy is not known [6]. However, the treatment of intussusceptions is always surgical and the resection may be necessary to some extent [19]. If contraindications of laparoscopy are not present, laparoscopic resection can be performed safely and should be considered for diagnosis and treatment for intussusception in ileocecal lesions in adults. The most important rule in treatment is the avoidance of tumor emboli spread during manipulation [20]. If the primary small bowel lymphoma is diagnosed before or at laparotomy, surgical resection should be preferred in order to limit the risk of serious complications, such as perforation, bleeding and obstruction. In early-stage patients, complete resection surgery is more advantageous in some reports [21-23].

After the surgery, not only can the histological diagnosis of the lymphomas be established but also accurate staging can be made. Chemotherapy and radiotherapy have been used either alone or in combination with surgery [24]. Some other studies do not support the need for radiotherapy in

intestinal B-cell lymphoma patients [6]. The results have not been evaluated adequately and the efficacy of these procedures is not known yet. Treatment strategies for primary small bowel lymphoma have been established but the debate about them persists [25]. However Radical tumor resectability had a highly significant effect on survival. In addition surgery in combination with chemotherapy was superior to any other treatment combination in localized disease and in disseminated cases [6]. Unfortunately, our patient refused post operative chemotherapy.

Conclusion

In small case series, few retrospective studies of small bowel lymphomas have been reported. In recent years some progress has been achieved in the diagnosis and treatment of gastric lymphomas, but the primary small bowel lymphomas are not well characterized and there is little information on their clinical diagnosis and management. However the treatment of ileal NHL revealed by intussusceptions in adults should always be surgical but preferably in combination with chemotherapy.

Conflict of interest

The authors declare that they have no competing interests.

Consent

Written informed consent was obtained for publication of this case report and accompanying images.

Authors' contribution

KIM is a surgeon who was drafting the manuscript and revising it critically for content. **HE**, **AA** and **AO** were involved in literature research and were major contributors in writing the manuscript. **KM**, **KA** were surgeons treating of the patient and were involved in revising the draft critically for content. **MB** and **ST** realized **US** and interpreted CT of the patient. All authors read and approved the final manuscript.

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