A rare case of chronic intussusception due to Non Hodgkin lymphoma

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Abstract

Chronic intussusception is defined as intussusception with a history of more than 14 days and is generally associated with a predisposing factor. We are reporting a rare case of chronic intussusception due to Non Hodgkin lymphoma of ileum, appendix, caecum and ascending colon presented as acute intestinal obstruction in emergency. Chronic Intussusception is rare in childhood due to Non Hodgkin lymphoma.

A five year male child presented with fever, pain abdomen, vomiting, diarrhoea and mass in right flank. Ultrasonography of the abdomen revealed a mass in ileao-caecal region with chronic intussusception which was confirmed on surgery. X ray of the abdomen showed dilated bowel loops.

It is very difficult to make diagnosis of intestinal lymphoma on pre-operative investigations. Patient presented with obstruction should be explored as surgery is the treatment of the choice. Diagnosis can be confirmed by histopathologically.

In conclusion, a high index of suspicion and appropriate investigations (USS, Barium enema and CT scan) can result in prompt diagnosis. In majority of children the diagnosis is made at laparotomy and surgery plays a pivotal role in the management. (Acta gastroenterol. belg., **2012**, 75, **42-44**).

Key words : obstruction, ultrasonography, intestine, tumour, surgery.

Introduction

Intussusception is invagination of a bowel segment, usually proximal, into a distal bowel segment. Intussusception is the most common cause of intestinal obstruction in children under two years of age (1). The classic triad of colicky abdominal pain, vomiting, red current bloody stools and a palpable mass occurs usually in acute intussusception (AI) but is rare in chronic intussusception (CI). Chronic ileocolic intussusception with intestinal obstruction secondary to neoplasm of the ileum, appendix, caecum, and ascending colon is extremely rare in childhood. The majority of cases in children are idiopathic.

CI occurs most typically in older children with a longer history and lower likelihood of successful hydrostatic barium enema reduction. Lymphomas of the gastrointestinal tract are the most common type of primary extranodal lymphomas, accounting for 5 to 10% of all Non Hodgkin Lymphoma (NHL) (1). It is necessary to diagnose properly a case of intussusception in the older children to prevent the failure of treatment (2). Primary lymphoma of the colon is rare and comprises less than 1% of large bowel malignancies. On extensive review of indexed literature only few cases of NHL presenting as ileocecocolic intussusception could be found (3). 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 children's.

Case report

A five year old boy presented with intermittent abdominal pain, vomiting, and an abdominal mass, on and off red recurrent bloody stools, constipation and diarrhoea since two weeks. The patient also had similar symptoms three months back. On examination the child was febrile, emaciated with signs of mild dehydration and tachycardia. A "sausage-shaped mass" in the right flank was felt on palpation. Right iliac region was empty (signe de Dance) and on per rectal examination, blood stained stool was found at tip of examining finger. Hemoglobin was 7.5 gm, the rest of investigations were within normal range.

Plain X-ray of the abdomen showed multiple air fluid levels in the erect and dilated ileal loops in the supine films. Ultrasonography (USG) confirmed small bowel loops forming a "target sign" which was diagnosed as chronic intussception (Fig. 1). There were significantly enlarged mesenteric nodes. The child was taken for emergency surgery in the view of intestinal obstruction after adequate resuscitation with intravenous fluids and antibiotics.

On exploratory laparotomy, there was a mass of 12 × 10 cm in size, at ileao-caecal region with invagination of the terminal ileum into the ascending colon extending upto 10-12 cm, which was non-reducible hence diagnosed as ileocolic intussusception. Mesenteric adenopathy with thickened mesentery were also present. Right hemicolectomy with ileotransverse anastomosis was performed. Multiple lymph node biopsies were taken. On the cut section of the specimen, there was ileocolic intussusception with a fleshy, yellowish tumour

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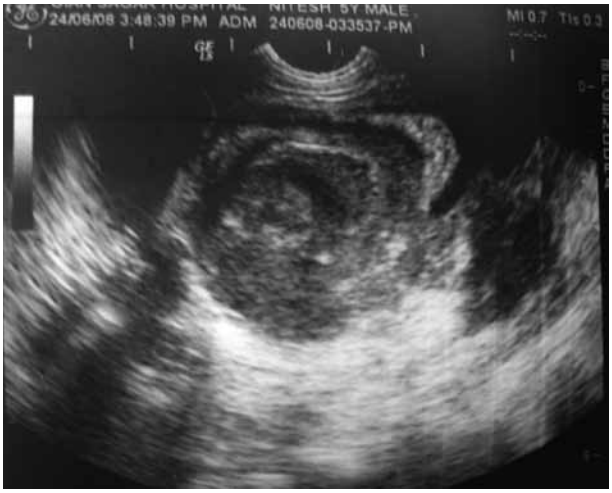


Fig. 1. — Ultrasonography of the abdomen revealed intussception as "target sign" with invagination of bowel into another with central mesenteric fat.

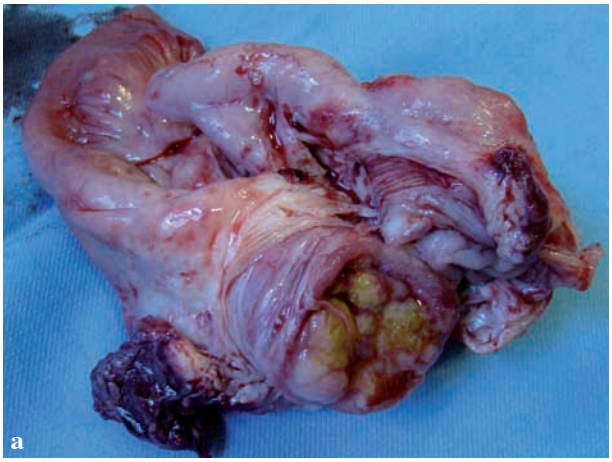


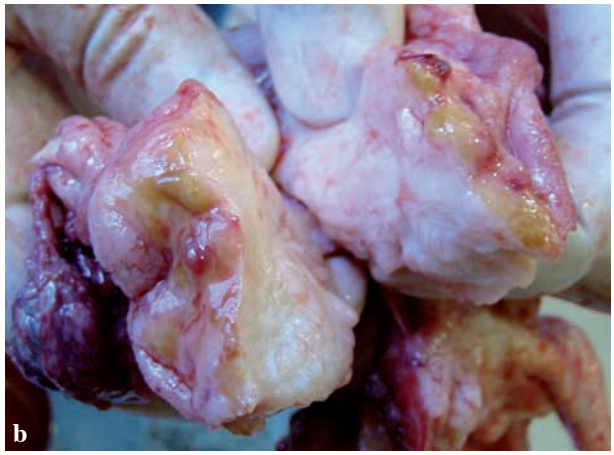
Fig. 2a. — Gross specimen showing ileocolic intussception with a tumour of fleshy yellowish colour and an enlarged appendix.

occupying the terminal ileum with multiple areas of haemorrhage and necrosis (Fig. 2 a-c).

Histopathology revealed malignant lymphoid cells within the submucosa, muscularis propria, and serosa of the terminal ileum, caecum and ascending colon extending upto adjacent fat diagnosed as NHL. The appendix also showed signs of infiltration by malignant cells (NHL cells). Lymph nodes showed reactive follicular hyperplasia. Post operative recovery was uneventful.

Discussion

Intussusception is a pathology in which telescoping of a proximal segment of bowel occurs into the lumen of the distal segment. The incidence of intussusception is 1.5-4 cases per 1000 live births, with a male-to-female ratio of 3:2 (4). Intussusception is a term derived from the Latin words "intuss" (within) and "suscipere" (to



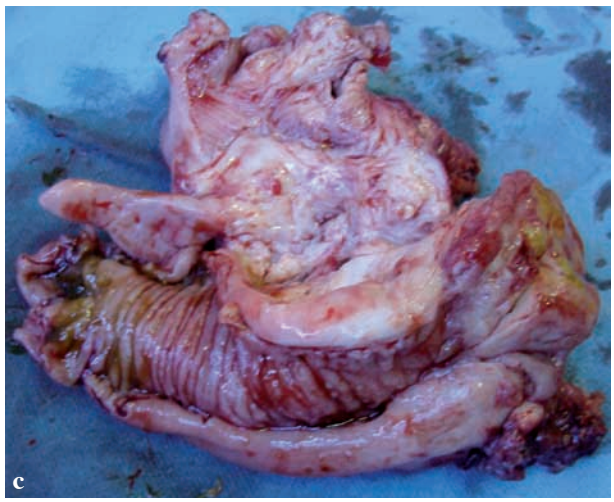


Fig. 2b, c. — On cut section of the bowel tumour and intestine along with the appendix.

receive). It often occurs around 1 year of age, with a peak incidence between 4 and 7 months (5). Chronic intussusception was first described by Rafinesque in 1878 in a group of children with intussusception who survived more than 14 days (4,6). Till 1995, 27 cases (mainly sporadic) were reported. Sometimes it is connected with NHL or in a sporadic case with Yersinia enterocolitica mesenteric adenitis. The sporadic form commonly presents with abdominal swelling as a large mesenteric, retroperitoneal or pelvic mass, tenderness, pain or fullness (7). Some patients present with symptoms of bowel obstruction secondary to ileao-caecal intussusception caused by tumor growth, obstruction or bleeding, mimicking acute appendicitis. Intussusceptions are classified according to location, with the most common classification system dividing intussusception into four categories : enteric, ileocolic, ileao-caecal, and colonic. Enteric and colonic intussusceptions are those that are confined to the small intestine and large intestine, respectively. Ileocolic intussusceptions are defined as those with a prolapse of the ileum through the ileocaecal valve into the colon and these constitute 15% of all intussusceptions (8).

Although 90% of the cases of intussusceptions in adults are associated with pathologic lesions of the bowel, the same is true for only 3% of cases in children under 1 year of age and 10% over 1 year of age (2,9). Intussusception is caused by a non-pathological lead point in more than 90% cases, whereas pathologic lead points are seen in less than 10% cases, i.e. Meckels diverticulum (75%), polyps, Peutz-Jeghers syndrome (16%), lymphoma & other tumors (3%) etc (4). However intussusception in an older child is usually associated with primary bowel pathology and malignant lymphoma is an occasional cause of non-strangulated intussusception (2, 4). The correlation between intussusception in older children and malignant lymphoma is not well documented in previous reports (3). CI is more commonly associated with a predisposing lesion and surgery is the treatment of choice in such cases. In case of the suspicion of malignant lymphoma of the bowel, it should be confirmed by USG, computed tomography (CT) scan and frozen sections.

An older child who does not respond to barium enema for reduction of intussusception should undergo surgery. Incomplete intussusception may be associated with diarrhoea and even diarrhoea can cause intussusception. Due to the high rate of unsuccessful hydrostatic reductions, all cases of CI should be treated surgically, as there is a high incidence of specific pathology which needs surgical intervention. Generally lymphoma arises in the lymphoid follicles of the submucosa of the bowel and proliferates into a localized large mass and polypoidal lesion and may invade the serosa to mesentery or beyond (10). Polypoidal lesions have better prognosis and infiltrating tumors with intestinal obstruction have poor prognosis. Spontaneous bowel perforation from lymphoma or during surgical manipulation increases the risk of peri-operative mortality. The extent of resection and operative technique depend upon the age of the patient, results of investigations (benign or malignant) and the length of the bowel involved. Surgery is the treatment of choice in intestinal obstruction due to malignancy and CI. Complete surgical excision together with chemotherapy using two or more cytotoxic agents provides the best results.

Conclusion

It is hoped that this article will draw attention to CI as a real clinical entity, and therefore be included in the differential diagnosis with a high index of suspicion in children who have prolonged abdominal pain with vomiting and weight loss. A high index of suspicion and appropriate investigations (USS, Barium enema and CT scan) can result in prompt diagnosis. In the majority of children the diagnosis is made at laparotomy and surgery plays a pivotal role in the management. Chemotherapy represents a cornerstone in the treatment of these patients and offers an excellent chance for long term disease free survival.

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