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Intussusception on a Small Bowel Gastrointestinal Stromal Tumor: A Rare Cause of Intestinal Obstruction in Adults

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Abstract **Case Report**

Acute intestinal intussusception is a rare surgical emergency in adults, especially when it involves an ileal gastrointestinal stromal tumor (GIST). An atypical clinical presentation, which may be associated with a partial or complete occlusive syndrome, a palpable abdominal mass or even melena, makes the use of radiological imaging essential, in particular CT scanning with contrast which allows the positive diagnosis of intestinal intussusception as well as identification of the organic lesion in question. Unlike children whose etiology is often idiopathic, in adults the incidence of malignant organic lesions is high. What makes the treatment of choice is complete surgical resection in one piece with healthy margins without intraoperative manual reduction. The diagnosis of GIST is confirmed by immunohistochemistry (CD117 – DOG1) or even a molecular biology search for mutations (KIT – PDGFRA). Adjuvant imatinib is essential when the risk of recurrence is intermediate or high. Rigorous monitoring to detect local and distant recurrences should not be neglected. We report the case of a 51-year-old patient admitted to the emergency room for an intestinal obstruction whose explorations concluded that it was an intestinal intussusception on an ileal GIST.

Keywords: Intestinal Intussusception, GIST, Surgical Resection, Case Report.

INTRODUCTION

Acute intestinal intussusception in adults, unlike children, is a rare manifestation occurring most often in an organic lesion (70 to 90% of cases), with a high incidence of malignancy. It represents 1 to 5% of etiologies of intestinal obstruction in adults [1]. Its progressive mode is usually chronic or subacute. It is rarely discovered in the face of an acute picture of intestinal obstruction or peritonitis [1, 2]. The small intestine represents approximately 30% of GISTs, with a lower risk of malignancy than the colon, as well as the exoluminal development making the complication of intussusception a rare situation [2, 3]. Here we report a rare case of ileoileal intussusceptions in a 51-year-old female patient who was successfully treated with segmental small bowel resection with anastomosis which was later found to be due to ileal GIST.

CASE REPORT

A 51-year-old woman lives in a rural area and has no significant medical history. With a history of intermittent abdominal pain for 03 months, becoming more and more intense and closer together for 15 days with the onset of bilious vomiting followed by a cessation of matter and gas as well as a deterioration in general condition made up of physical asthenia and anorexia.

Referred from another hospital, upon admission to the emergency room the physical examination found a conscious patient, bedridden, slightly discolored conjunctiva, tachycardia at 110bpm, the abdomen was distended with palpation of a tender and mobile mass at the fossa level right iliac. The biological assessment shows anemia at 9 g/dL, hypokalemia at 3 mEq/L as well as an elevation of CRP to 73 mg/L. An abdominal X-ray was performed, showing hydro-aeric levels, and investigations were completed by abdominal CT scan with iodinated contrast injection, showing irregular parietal thickening of an ileal loop complicated by ileo-ileal intussusception with an upstream occlusive syndrome, of probable lymphomatous etiology., of which a lymphomatous etiology is probable.

After preoperative preparation including nasogastric decompression, rehydration with potassium supplementation and antibiotic prophylaxis, the patient

was admitted to the operating room the same day. A median laparotomy straddling the umbilicus reveals significant small bowel distension upstream of an ileo-ileal intussusception at the level of the right iliac fossa located one meter from the ileo-caecal valve, we note whitish parietal nodules of the invaginated handle. No attempt has been made to manually reduce the intussusception tube. The operation consisted of a segmental small bowel resection encasing the intussusception coil with creation of a manual end-to-end ileo-ileal anastomosis, without any attempt at intraoperative manual reduction. The early postoperative course was simple with discharge home on POD5.

The anatomopathological and immunohistochemical studies of the surgical specimen concluded that a polypoid mass of 06*05 cm was in favor of a panparietal mesenchymal tumor not expressing CD117, a complement of molecular study made it possible to retain the diagnosis of gastrointestinal stromal tumor with intermediate risk of recurrence. The decision of the multidisciplinary consultation meeting was to put the patient on Imatinib with monitoring in the oncology department.

Currently 6 months after surgery, the patient is doing perfectly well with clinical-radiological control without abnormalities.

DISCUSSION

Intussusception refers to the telescoping of the proximal segment into the lumen of the distal segment of the intestine causing obstruction, ischemic injury, and ultimately gangrene and perforation [2, 3]. Intussusception usually occurs during the fifth and ninth months of life and is rare in adults, accounting for 1% of all intestinal obstructions and only 0.003–0.02% among all hospitalized patients [4, 5]. In adults, an organic cause is found in 70 to 90% of cases while 8 to 20% are idiopathic. However, intestinal intussusception is primary in children in 90% of cases [1-6]. This organic cause may correspond to malignant tumors representing the primary etiology of intussusceptions in adults especially in the colon, while they are secondary to a benign lesion (especially in the small intestine) in 25% of cases and 10% idiopathic. These lesions are represented by stromal tumors, lipomas, polyps, lymphadenopathy, digestive thickenings. Melanoma, adenocarcinoma, etc [1]. Any intraluminal lesion that alters normal peristaltic activity, producing an area of constriction and relaxation sequence, thereby telescoping the digestive loop across the distal lumen of the intestine [7]. Acute intestinal intussusception in a small bowel GIST is rare, like the case of our patient.

GISTs are rare gastrointestinal tumors that originate from interstitial Cajal cells or precursor stem cells and are observed particularly over the age of 40 [2, 3]. They represent less than 0.2% of all gastrointestinal tumors and only 0.04% of small intestinal malignancies which may occur as a primary cause in adult intestinal intussusceptions [2]. They can occur anywhere along the gastrointestinal tract, with 60–70% in the stomach and 20–25% in the small intestine [8]. Intussusception confined to the small intestine is rare and accounts for approximately 10% of all intussusceptions in adults, with some studies showing a figure as low as 1% [4].

Patients with GIST generally present with symptoms such as melena, partial or complete intestinal obstruction, such as abdominal pain, nausea, vomiting and early satiety depending on the site of the tumor [3].

The diagnosis of ileo-ileal intussusceptions can be simple in the presence of a palpable abdominal mass related to the intussusception sausage. However, the rarity of this pathology in adults as well as its atypical clinical presentations are the main causes of preoperative diagnostic errors or diagnostic delays [2, 3]. Our patient presented a palpable mass in the right iliac fossa with an occlusive syndrome.

Abdominal ultrasound has high sensitivity and specificity in the diagnosis of intussusceptions, while abdominal radiographs are important to exclude obstruction in an emergency situation [1-6]. Abdominal CT with injection of the contrast product remains the examination of choice in this situation, especially in cases of emergency with occlusive syndrome, allowing reliable identification of intussusception and increasing the sensitivity of the diagnosis which can reach 90% with a specificity of 100% in adults [1]. It makes it possible to diagnose the obstructive syndrome, its mechanism, in this case intussusception, its precise location and to show its cause. It can detect an organic cause in 71% of cases. The two classic images are the “sandwich” image in longitudinal section showing the head of the intussusception and the “roundel” image in cross section showing the sausage of the intussusception. The use of MRI and endoscopy should be discussed according to needs [2].

However, preoperative diagnosis is often difficult due to the nonspecific presentation of the tumor, hence the recourse, according to some authors, to diagnostic laparoscopy [2].

The treatment of intussusception in adults is always surgical and the intervention of choice remains resections according to the rules of oncological surgery given the high incidence of malignancy [2-5]. It still remains difficult to differentiate a malignant etiology from a benign etiology preoperatively and intraoperatively, thus presenting a risk of transperitoneal seeding following tumor breakout during the handling of friable malignant tissues, recommending against intraoperative reduction of intussusceptions [1-5]. Intraoperative histopathology can help determine the extent of surgical resection in advanced settings [9]. Thus, resection is the treatment of choice in adults with

intussusceptions without any attempt at manual reduction [1-3].

The treatment of choice for localized GIST is complete surgical resection. Routine removal of lymph nodes is generally not necessary because lymph node metastases are rare [2].

The histopathological diagnosis of GIST must be confirmed by an immunohistochemical study, which notably makes it possible to demonstrate expression of c-KIT (CD117) by tumor cells in 95% of cases [10, 11]. The systematic use of a second antibody, DOG-1, particularly in cases of negative c-KIT is recommended, with also a positivity in more than 95% of cases. Determination of the mitotic index (on $5mm^2$) is fundamental to assess the risk of recurrence. Note that Ki-67 has no demonstrated value in GIST.

The search for mutations in the KIT and PDGFRA genes using a molecular biology technique, in addition to allowing the diagnosis to be confirmed in difficult cases, is now part of current practice in the management of GIST. Indeed, the type of mutation has an influence on the prognosis and the effectiveness of treatment in the adjuvant and metastatic situation. Genotyping of GISTs is recommended with the exception of GISTs with a very low risk of recurrence [10, 11].

As there is a possibility of KIT or PDGFRA mutation in c-kit negative GISTs, complete resection and adjuvant tyrosine kinase inhibitor imatinib should be considered to significantly reduce metastasis and the risk of local recurrence of GISTs [2, 3]. Our management consisted of a segmental small bowel resection involving the intussusception coil with creation of an ileo-ileal anastomosis without intraoperative reduction as well as an adjuvant treatment based on Imatinib taking into account the size of the tumor and the risk of recidivism.

Even after complete surgical resection, recurrence of GISTs may occur, so the patient needs regular follow-up and investigations to detect local and distant recurrences [3].

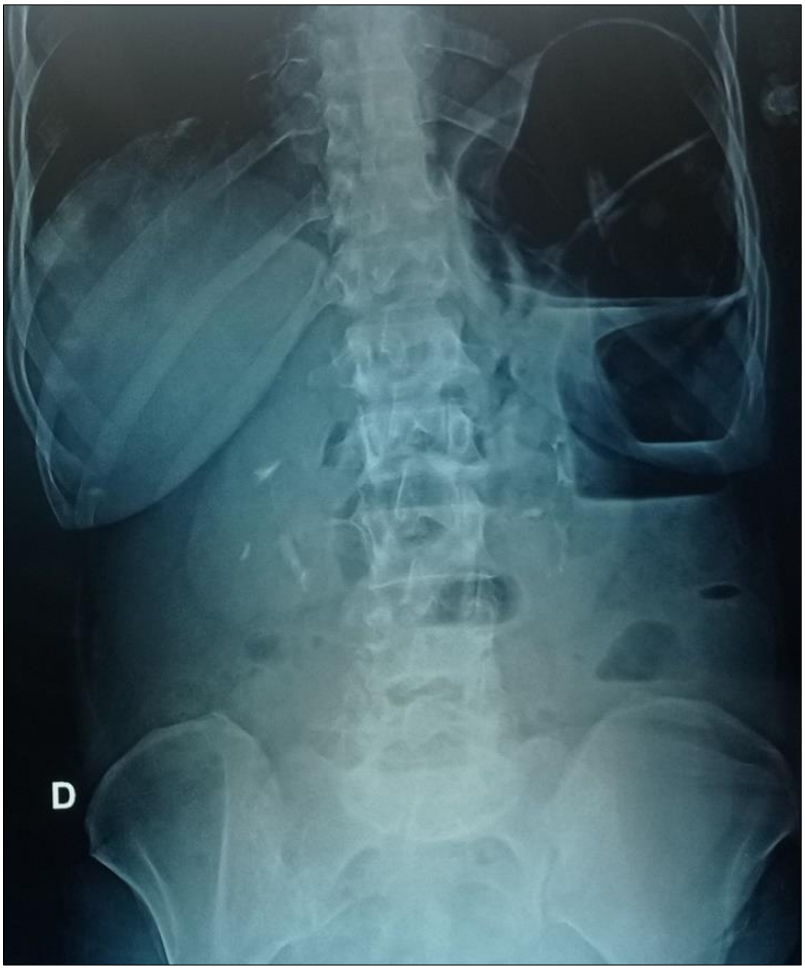

Figure 1: Abdominal x-ray showing air-fluid levels.

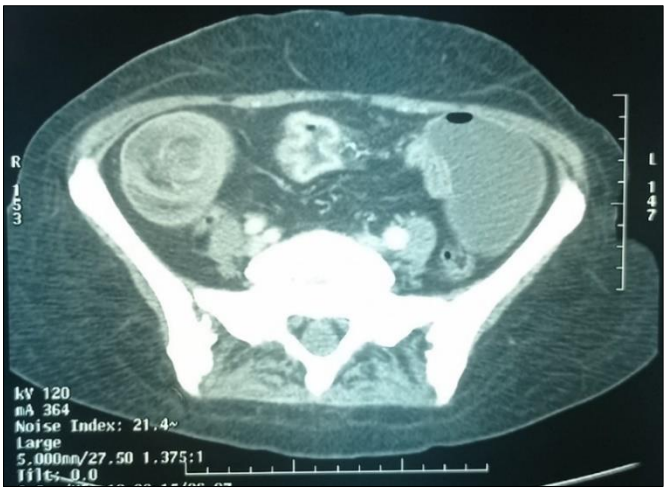

Figure 2: Axial sections of the abdominal CT scan showing irregular parietal thickening of an ileal loop with a cockade appearance suggesting an ileo-ileal intussusception.

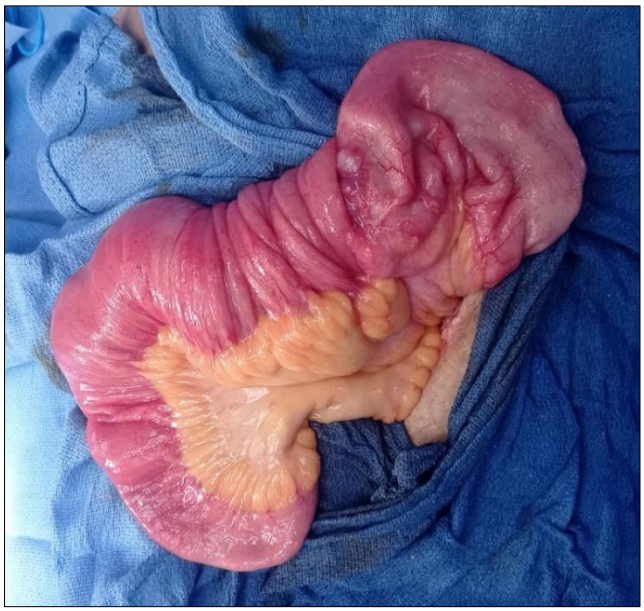

Figure 3: Operative findings, ileo-ileal intussusception of the parietal whitish nodule of the invaginated loop.

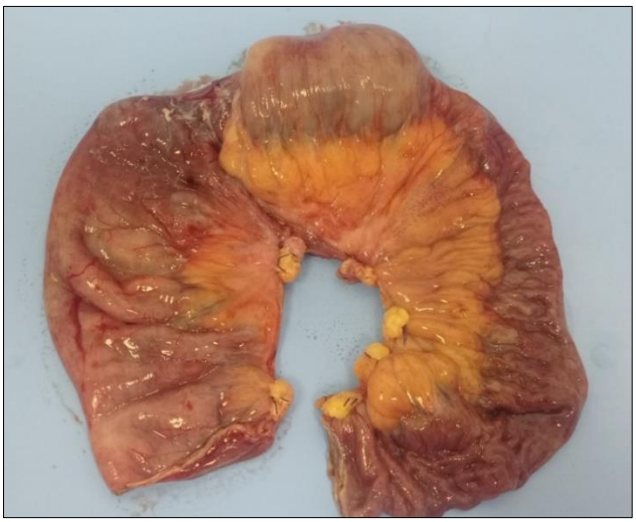

Figure 4: Operating specimen, segmental small bowel resection containing the intussusception coil.

CONCLUSION

Ileoileal intussusception due to GIST is a rare clinical entity in adults. Its preoperative diagnosis is quite difficult due to the rarity of the disease and its nonspecific presentation. Computed tomography represents the imaging modality of choice allowing a definitive positive and etiological diagnosis. Surgical resection is the standard treatment. Depending on the risk of recurrence defined by histopathological or even molecular studies, adjuvant Imatinib with regular monitoring allows the reduction of local and distant recurrences of GIST.

Conflicts of Interest: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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