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CASE REPORT

Intestinal duplication in adulthood: A rare entity, difficult to diagnose

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Abstract

Duplications of the alimentary tract (ATD) are rare congenital anomalies often found early in life. They may occur anywhere in the intestinal tract but the ileum is the most frequently affected site. Clinical presentation of ATD in adults is variable and because these lesions occur so infrequently they are rarely suspected. In the present report we describe a case of ileal duplication in a 61-year-old patient with Crohn's disease. Despite various radiological investigations and medical consultations, the diagnosis was only made on the surgical specimen.

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Key words: Intestinal duplication; Adulthood; Intestinal obstruction; Surgical resection; Abdominal pain

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INTRODUCTION

Duplications of the alimentary tract (ATD) are rare congenital anomalies. Eighty percent of ATD are diagnosed in children before the age of 2 years. They may occur anywhere in the intestinal tract but the ileum is the most frequent affected site[1-3].

Due to the infrequency of ATD and its major relevance in the pediatric population, the analyses of patient characteristics and clinical manifestations in the adult are limited.

In the present report we describe a case of ileal duplication in a 61-year-old patient with Crohn's disease.

CASE REPORT

A 61-year-old Caucasian patient with a 5-mo history of intermittent diffuse abdominal pain associated with nausea, vomiting and weight loss was referred to our Gastroenterology Unit with a diagnosis of ileal Crohn's disease. A small bowel follow through (SBFT) showed an ileal stricture associated with dilation above a stenosis and a suspected entero-enteric fistula at about 30 cm from the ileocecal valve (Figure 1). However, no radiological evidence of mucosal lesions, including ulcers, was detectable. Steroid treatment was given, followed by incomplete remission.

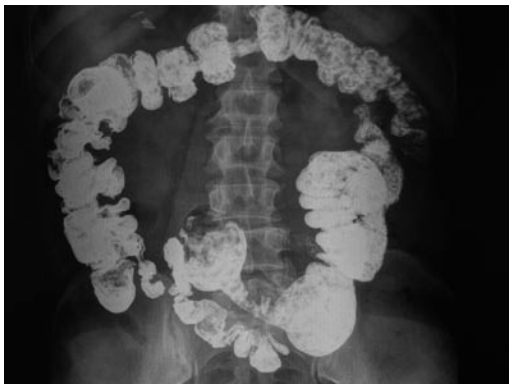

Figure 1 Small bowel follow through shows an ileal stricture associated with dilation above stenosis and suspected entero-enteric fistula at about 30 cm from the ileo-cecal valve. No conclusive diagnosis was made on the basis of this examination.

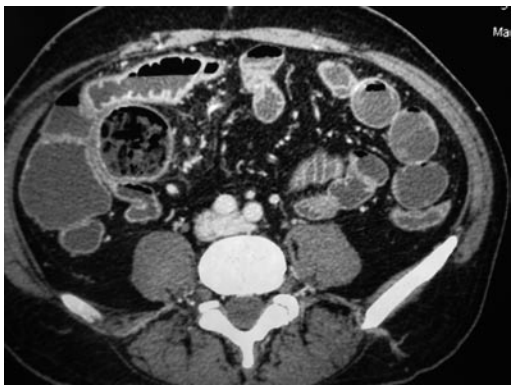

Figure 2 Entero-computed tomography showing a marked ileal dilatation, with no evidence of mucosal lesions or fistulae.

Clinical examination and blood tests at admission were normal. The patient had no family history of inflammatory bowel disease (IBD) and there were no abdominal masses, cutaneous fistulas or other stigmata of Crohn's disease. A Small Bowel Contrast Ultrasonography (SICUS) was performed that showed findings comparable to SBFT. Oral budesonide (9 mg/d) was given with temporary and partial benefit. Entero-computed tomography (CT) scan confirmed the ileal stenosis associated with dilation above the stricture but it also visualized a blind loop of the intestine close to the stenosis (resembling a diverticulum) and enlarged mesenteric lymph-nodes (Figure 2). At this point, a differential diagnosis between CD and intestinal lymphoma was made in a symptomatic patient, giving an indication for an explorative laparotomy.

At surgery, an inflammatory mass was found in the right iliac fossa. The terminal and pre-terminal ileum, the cecum and the great omentum were involved. Tight adhesions were found between the above mentioned structures and both the terminal ileum and regional mesentery. The intestine at this level was thickened and a pre-stenotic dilation was present. A standard ileo-cecal resection was performed and the surgeon described the intra-operative findings compatible with an inflammatory mass of unknown origin. The patient made an uneventful recovery and was discharged home within a week.

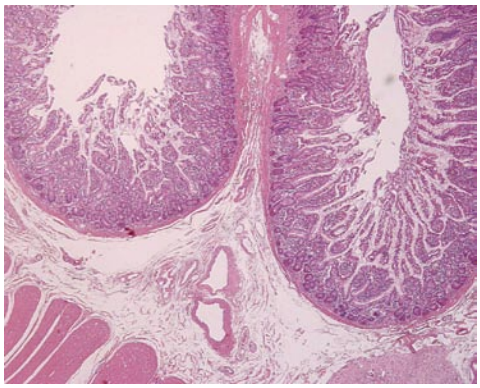

Figure 3 Histological section of the intestinal duplication at low magnification, showing the wall structure including the mucosa, the submucosa and the inner circular layer of the muscolaris própria (hematoxilin-eosin, original magnification 20 ×).

Macroscopic examination showed a tubular structure communicating with the ileal lumen that measured 5.5 cm × 2.5 cm × 2 cm. The cut surface was morphologically indistinguishable from the normal ileal wall. Light microscopy (Figure 3) highlighted the four layered organization of the ATD wall, including a mucosa with an intestinal-type epithelial lining and a muscularis propria, with an inner circular and outer longitudinal smooth muscle layers. A myenteric plexus was present between the two muscle coats.

The mucosa showed patchy features of ischemic injury. A diagnosis of ileal duplication was finally made.

DISCUSSION

ATD are congenital anomalies of the intestine, first described by Fitz[4]. They can be spherical or tubular and can be attached or adherent to the ATD. These conditions are rare (1/10000 live births), usually encountered in the ileum[1,2] and the vast majority are found in infants[5,6]. The differential diagnosis is with mesenteric cysts and true and false diverticula. However, a duplication shares a portion of its wall with the adjacent small intestine, usually sharing a common blood supply. The epithelial lining is always of some part of the ATD and may include heterotopic gastric mucosa. Malignant degeneration has been described in the adult series[7].

According to the review published by Johnson *et al*[8] in 1994, cancer was found in 3 (23%) of 13 reported cases of ileal duplications in adults (2 adenocarcinoma and 1 squamous cell carcinoma). This evidence of epithelial instability might suggest a tendency toward malignant transformation in long standing duplications. This also supports complete resection of the duplication as the most appropriate method of treatment.

Clinical presentation of ATD in adults is variable and, because these lesions occur so infrequently, they are not suspected. A palpable mass can be found in approximate-

ly one half of patients; abdominal pain is often present but the most common clinical presentations include intestinal obstruction and bleeding[1,9-11]. It is worthwhile to highlight that the clinical presentation is strictly related to the site and type of ATD. In cases of ATD of the hindgut, the diagnosis is often made within the first years of life and the most frequent symptom is biliary vomits. These malformations are usually cystic and localized on the mesenteric border of the first or second duodenum. In the jejunum, the most frequent aspect includes a tubular duplication with a common lumen, whilst in the ileum ADT can resemble a diverticulum. Ileal duplication affecting the distal part of the intestine should be distinguished from a Meckel's diverticulum, even though this is present on the anti-mesenteric border of the intestine. Complications of ATD include volvulus, invagination, bleeding, perforation and malignancy.

Twenty-seven cases of ileal duplications in adults are described in the world literature in over 100 years. In one of these cases, the correct diagnosis was made preoperatively. In this case, clinical presentation and pre-operative studies supported a diagnosis of complicated CD. Biopsies were not taken because it was not possible to enter the ileo-cecal valve during diagnostic colonoscopy and the rest of colonic mucosa was normal. Laparotomy is also often indicated in these settings to make a differential diagnosis[12]. Abdominal scans such as SICUS, CT or MRI and conventional contrast x-ray studies are useful tools to detect ATD. The diagnostic problems arise from the extreme rarity of this entity in the adult population.

We hereby describe a case of an adult patient who underwent various radiological studies and was referred to different physicians during the year before the correct diagnosis was made. The patient had 3 previous admissions to A&E and was on oral steroids when referred to our Gastroenterology Unit. ATD was not suspected and the diagnosis was made on the surgical specimen.

In conclusion, ATD are congenital abnormalities that can arise at any level from the mouth to the anus. They are rare and often found early in life. A minority of cases may remain undiscovered until adulthood when they may give rise to different symptoms, depending on the location. The ileum is the most frequent affected site and abdominal pain is the most referred symptom. Diagnosis is difficult due to the rarity of this entity. Symptoms are not specific and intestinal duplication is not considered in differential diagnoses. We believe therefore that it is useful to report new cases and to review the most relevant aspects of this entity. Surgical correction is the treatment of choice and, in the adult, resection of the entire duplication should be undertaken due to the reported incidence of malignancy.

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