The Italian Journal of Surgical Sciences
Vol. 19, No. 4 : 395-398, 1989

Intestinal perforation caused by non specific idiopathic ulcer of the small intestine. A case report

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Key Words: small intestine perforation, non specific idiopathic ulcer

A case of non specific idiopathic ulcer of the small intestine in a 79-year-old patient is reported. On admission he presented with acute abdomen and was submitted to wide intestinal resection. The features of this rare disease (only 370 cases described until 1987), which has an undefined etiopathogenesis, severe symptoms, and poses diagnostic problems, are discussed.

Introduction

This paper reports a case of non specific idiopathic ulcer of the small intestine, an extremely rare disease [3, 6, 13, 17] of which according to M.R. Watson (1963) [19], only about 170 cases had been reported in the literature after the first autopsy case described by Matthew Baillie in 1795 [4]. More recently (1987) R. Marchetto [14] has stated that 370 cases of primary isolated ulcers of the small intestine have been observed up to now.

Case Report

A 79-year-old female patient was admitted to our department because of intense abdominal pain and continuous remittent fever which did not respond to medical treatment. The patient had been on oral theophylline treatment for hypertension and respiratory distress for about a year before admission.

Physical examination revealed tenderness in the right iliac fossa. Gas evacuation was present. Direct X-ray did not evidence any significant alterations. Neutrophil leukocitosis was present.

The patient underwent surgery after diagnosis of circumscribed peritonitis probably induced by acute appendicitis. Laparotomy revealed a septic focus involving the omentum, ileal loops and cecum. This was due to rupture of one of the intestinal loops which caused marked fecal leakage. A perforating ulcer about 1.5 cm. in diameter was observed just above the rupture.

Moderate vascular impairment seemed to be present in the distal intestinal loops, cecum and ascending colon. The mesentery of the distal loops was slightly thickened.

Viscerolysis was performed and extensive intestinal resection was considered. However a section of the intestine about 10 cm. from the cecal ileal valve and examination of the surface of the loop suggested removal of the

cecum and part of the ascending colon together with the affected loops in order to perform anastomosis to healthy tissues.

The colon and ileal stumps were sutured with a stapler and side-to-side ilio- transversocolic anostomosis was carried out with an extramucosal monolayer suture.

Because of the presence of a large number of stones in the gallbladder cholecystectomy was carried out. A drain was positioned in the abdominal cavity.

Histological examination revealed two acute, perforated non specific ulcers, one circumscribed and the other surrounding the whole circumference of the intestine.

Marked fibrin and leukocyte exudation was present in the perivisceral fat where there were signs of recent thrombosis, vascular congestion and bleeding. The mesenteric lymph nodes presented histiocytosis of the sinuses.

The mucosa was hypotrophic with marked granulocyte infiltration with a great number of eosinophils in the lamina propria. Numerous lipofuscin stained macrophages were observed.

The patient's postoperative course was characterized by diuretic disorders related to a pre-existing renal insufficiency and by hypertensive crises accompanied by respiratory distress.

The patient recovered and was discharged from hospital on day 45 after admission.

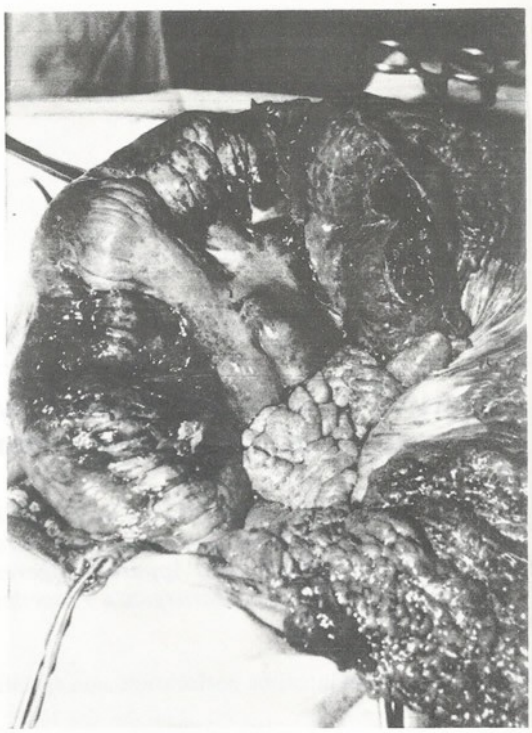

Fig. 1. *Complete dissection of the terminal ileum due to perforation of ulcer along the whole circumference of the intestine, is clearly shown.*

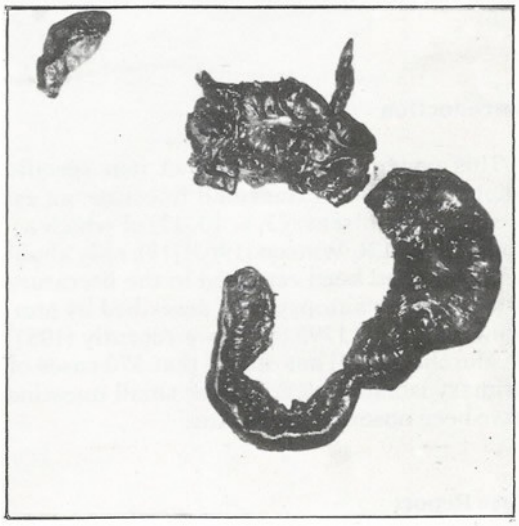

Fig. 2. *Ileum, cecum and a portion of ascending colon are excised. Note the ulcerative lesions.*

Discussion

Non specific idiopathic ulcers of the small intestine are usually located in the distal loops of the ileum with no predisposition for age or sex. The pathogenesis is uncertain [18]. It seems to be a specific nosographic condition even if there are vague hypotheses linking it to necrotizing enteritis and Crohn's disease [1, 11].

This idiopathic ulcer could be caused by iatrogenic or vascular factor.

The former could be provoked by administration of thiazide diuretics especially in combination with potassium chloride [12].

Baker et al. (1964) [2] first reported the side effects of the gastro-resistant form of this drug and the clinical observations were confirmed in the experimental animal and in vitro studies; the ulcerogenous effect of this drug could be due to elevated, rapid K concentration in the tissues once the enteric juice dissolves the gastro-resistant capsule. There are reports in the literature regarding the possible role of theophylline in determining

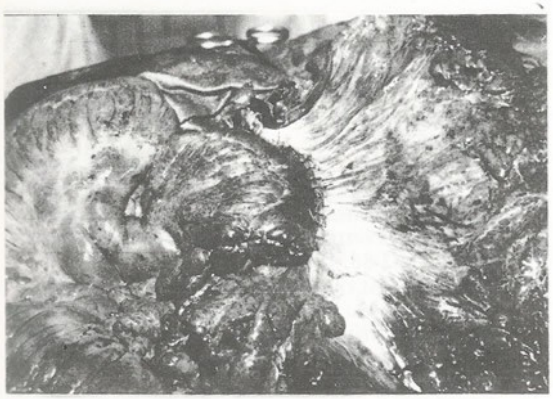

Fig. 3. *Side-to-side ilio-tranversocolic anastomosis with extramucosal monolayer suture.*

necrotizing enteritis [7, 15]. This drug is used in newborns with respiratory problems related to premature birth.

Although there is discordance concerning the correlation between drug and necrosis, it must be kept in mind when studying the etiology of drug-induced intestinal disorders. Thus, it is important to remember that our patient was previously treated with theophylline.

The possible ischemic nature of the disease is based on the well-known fact that marked but not necrotizing ischemia of the small intestine provokes severe mucosal degeneration [8, 9]. However, the muscular and serous tunicas may not suffer excessively from the vascular deficit because they possess a well-developed and almost autonomous intramural circulation [5].

The not infrequent circumferencial aspect of the ulcerations [10] could be explained by this ischemic impairment which could provoke complete rupture of the intestine as seen if the perforation involves the whole circumference of the intestine.

Angiographic and anatomical studies [8] demonstrated that circulation in the terminal ileal loops is more difficult than in all others because the distal ileum is vascularized by a single marginal arch which gives rise to long, scattered and often thin terminal vessels [16]. The eosinophil granulocyte parvicellular infiltration present in our case is a common finding and suggested the possible allergic origin of the ulcer, as in Arthus's phenomenon. However, it is evident that ulcers have a multifactorial origin.

The clinical picture of non specific idiopathic ulcer of the small intestine is not clear and consists of dyspeptic disorders and mild abdominal pain. Symptoms are more manifest when chronic (anemia, constipation) complications or acute (bleeding, perforation, intestinal occlusion) occur.

Bleeding poses a difficult diagnostic problem which requires selective arteriographic examinations.

The positioning of the catheter in the afferent vessel at the ulceration site can facilitate surgical detection of the intestinal segment involved [10].

Perforation is frequently covered and this can also complicate differential diagnosis since there is no free air in the peritoneum.

Intestinal obstruction caused by the healing and rupture of the ulcer is often mistaken for Crohn's disease.

Surgery can easily cure this disease. Its real interest lies in its rarity, undefined etiopathogenesis, diagnostic difficulties and severity of its complications.

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Italian Abstract – Un caso clinico di ulcera idiopatica non specifica.

Gli autori descrivono un caso di ulcera idiopatica non specifica del piccolo intestino in una paziente di 79 anni giunta alla loro osservazione con il quadro clinico di addome acuto ed operata di ampia resezione intestinale.

Vengono discusse le caratteristiche di questa rara malattia (solo 370 casi descritti in letteratura fino al 1987) che presenta notevole interesse per la sua etiopatogenesi oscura, la severità dei sintomi, la difficoltà nella diagnosi.

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