Case report

A rare location of Crohn's disease: the nasal mucosa

Rinaldo Pellicano[1], R. Sostegni[2], C. Sguazzini[1], S. Reggiani[1], M. Astegiano[1]

[1] Department of Gastro-Hepatology, Molinette Hospital, Turin; [2] Gastroenterology Unit, Mauriziano Hospital, Turin

Abstract. Crohn's disease (CD) is a chronic inflammatory disease that affects both the small and large intestine in approximately 40% of cases, solely the ileum or the colon in 30% and 25%, respectively. The remaining locations of the gastrointestinal (GI) tract are involved in percentages ranging between 0.5 and 5%. The appearance of the disease outside the GI tract is an exceptional event. In the present case, the authors report the history of a male patient suffering from CD involvement of almost the entire digestive system plus the nasal mucosa. This latter event emerged after repeated episodes of epistaxis, the demonstrations of histologic nasal features similar to those of intestinal CD, and the remission after treatment with beclomethasone. Since in literature less than a decade of cases of nasal location of CD was described, it is of prime importance to highlight that in CD patients, the occurrence of repeated episodes of epistaxis should prompt a consideration in the differential diagnosis of nasal location of the disease. (www.actabiomedica.it)

Key words: Crohn's disease, beclomethasone, nose

Introduction

Crohn's disease (CD) is a chronic inflammatory disease of unknown etiology that affects mainly the bowel (1), but every organ of the digestive system might be involved (2). The location outside the gastrointestinal (GI) tract is considered an exceptional event.

Case report

E. P., male, born in 1959, presented with a remote pathological anamnesis of facial paralysis *a frigore* which occurred at the age of 27 years, and hypertensive crises which lasted for months, defined as neurovegetatives, documented in the following 3 years.

In 1990, following a persistent and intense epigastric pain, the patient underwent upper gastrointestinal endoscopy (EGD), during which gastric ulcer was diagnosed. The lesion was treated with H2-antagonist drugs at full dose for 6 weeks with no symptomatic benefit, and then with proton pump inhibitors at full dose for 4 weeks, but after that period, the ulcer did not show any regression. Therefore, the addition of sucralfate was proposed. A few months later, after digestive hemorrhage from the same location (Fig. 1), the patient underwent gastrectomy with Roux –en-Y anastomosis. Histology diagnosed "gastric ulcer with periulcerative gastritis, chronic granulomatous gastritis and reactive lymph nodes".

In 1991, following the onset of diarrhea with relevant anemia and rectal bleeding, E. P. was hospitalized again. At colonoscopy, "stenosis for an extension of 15 cm associated with aphthous ulcers" was detected in the cecum and ascending colon as well as a "modest narrowing" of distal ileum. He was discharged with a diagnosis of CD localized in the stomach, ileum and colon and he was sent to our attention.

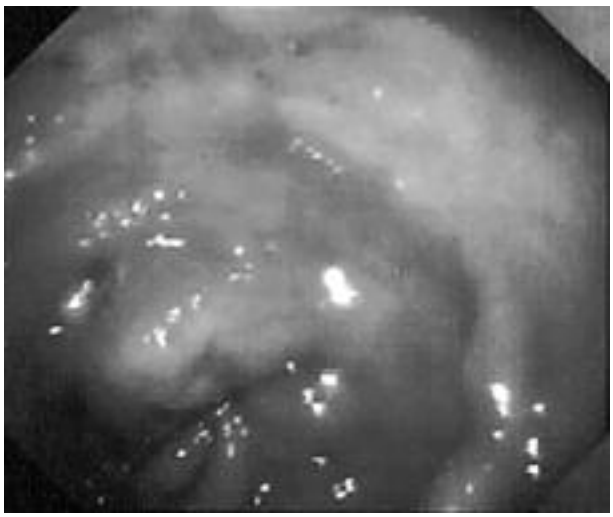

Figure 1. Bleeding gastric ulcer of the described patient suffering from Crohn's disease involving the entire gastrointestinal tract

The following year, due to CD relapse, a treatment with oral corticosteroids tapering (prednisone, starting with 50 mg/day and reducing by 5 mg every 10 days) was started. At the end of this treatment, a long-term maintenance therapy with oral mesalazine (800 mg/day) was performed. After 1 year, following an acute abdomen due to a tamponade perforation of the last ileal loop with a pericecal abscess, E. P. underwent, in emergency, "right colectomy plus resection of 80 cm of terminal ileum and ileal-transverse end-to-side isoperistaltic anastomosis". Histologic examination revealed a framework of "multiple ulcers, transmural inflammatory infiltration and sclerosis, multinodular chronic lymphadenitis with granulomatosis features. Thereafter, the patient had no symptomatic benefit and showed persistent diarrhea, abdominal pain mainly localized in the lower regions and polyarthralgia of the large joints, resulting in oral corticosteroid therapy (prednisone, starting with 50 mg/day, reducing by 5 mg every 10 days and maintaining the dose of 10 mg/day for 7 months) associated with cycles of ciprofloxacin being started again.

In association with these events, epistaxis appeared which persisted for over 5 months, with episodes of clinical relevance followed by anemia. Analysis of blood samples showed normal blood coagulation parameters, and instrumental examinations (X-ray of the paranasals sinuses and computed tomography of the facial bones) revealed a well-defined mucosal thickening at the base of the left maxillary sinus where biopsies were performed. These showed "mucosa covered by stratified squamous epithelium, site of acanthotic hyperplasia and hyperkeratosis, underlying chorion surface infiltrated by elements of nonspecific inflammation, evidence of granulomatosis features". The diagnosis was nasal localization of CD. In June 1994, following the development of anorectal sepsis, the patient went to Saint Mark's Hospital in London where an abscess was drained and 4 setons were placed to facilitate drainage through a fistula. On this occasion, the diagnosis of nasal CD was confirmed and treatment with beclomethasone spray (200 mcg/ml, 1 ml per session 3 times a day) was introduced. This was continued for 5 months with good control of ENT manifestations.

After an intermittent pattern of disease during the years 1995-1998, the patient came to our attention for a control visit in March 1999 with increased weight, 3 normochromic fecal discharges daily and without episodes of epistaxis. During this period, he had undergone continuous treatment with mesalazine 250 mg 6 times daily. However, in the course of the same year, he was hospitalized for pneumonia and intravenously treated with cephalosporins.

In subsequent years, nasal and intestinal manifestations of CD followed an intermittent trend with use on demand of corticosteroids and monthly administration of antibiotics.

In April 2003, the patient was urgently admitted to the hospital for jejunal perforation and underwent "jejunal resection and end-to-end anastomosis in a double layer of the first jejunal loop and of the loop of Roux –en-Y anastomosis with a fistula; bowel segment isolation and resection of 35 cm of terminal ileum plus the anastomosis and 5 cm of transverse colon; protective transient ileostomy on the last ileal loop". Histologic examination described "chronic transmural inflammation, extensive ulcerations with areas of pyloric metaplasia, submucosal edema, peritonitis, reactive hyperplasia in perigranular lymph nodes". A few days later, following hematoma and perianastomotic ischemic colitis, EP underwent "resection of the last 30 cm of terminal ileum and ileostomy in the right iliac

region". In the following days, he had respiratory failure of infectious origin (from bronchial aspirate *Staphylococcus aureus*, *Corynebacterium species*, *Escherichia coli* and *Candida albicans* were isolated) resistant to targeted and broad-spectrum antibiotics and antifungal treatments. The progressive deterioration induced shock, cardiac arrest and exitus.

Conclusions

In the population of Northern Italy, the incidence of CD is 3.4/100.000 inhabitants/year (3). At the beginning, the concurrent involvement of small and large intestine affects approximately 40% of cases, while the exclusive ileal involvement is equivalent to 30% and that of colon to 25% of patients, respectively. The remaining locations of the GI tract are involved in percentages ranging between 0.5 and 5% of cases (4).

The early lesion of CD is the aphthous ulcer endoscopically recognizable like minute erosion with white bottom and hyperemic margins, which tend to deepen in the mucosa with longitudinal fissures and appearance "to cobblestone" mucosa. The granulomas are the only feature of certain diagnosis and consist of epithelioid cells, occasional Langhans giant cells and lymphocytic mantle. However, granulomas are found in only 50% of cases, and in the remaining ones, diagnosis is based on the histological evidence of nonspecific transmural inflammation combined with clinical data (2).

Although in literature cases of location of CD in almost all the digestive system are described, descriptions of nasal involvement are rare. In a literature review, published in 2004, only 4 descriptions of nasal localization of CD were given (5). In these cases, the diagnosis occurred after episodes of sinusitis with polypoid features, nasal congestion or nasal stenosis (5).

The involvement of nasal mucosa in CD in E. P. is well documented, considering the identical diagnosis in 2 different centers (in Italy and in UK) and the success in the symptomatic control through the use of beclomethasone. This description is even more relevant when one considers that this case is unique in our outpatient center, in which between 10,000 and 12,000 visits are yearly perfomed (6) and of which approximately 2400 for inflammatory bowel disease (7).

The message from this case report is that in CD patients, the occurrence of repeated episodes of epistaxis should prompt a consideration in the differential diagnosis of nasal location of CD.

References

1. Abraham C, Cho JH. Inflammatory bowel disease. *N Engl J Med* 2009; 361: 2066-78.
2. Lichtenstein GR, Hanauer SB, Sandborn WJ and The Practice Committee of the American College of Gastroenterology. Management of Chron's disease in adults. *Am J Gastroenterol* 2009; 104: 465-83.
3. Ranzi T, Bodini P, Zambelli A, et al. Epidemiological aspects of inflammatory bowel disease in a north Italian population: a 4-year prospective study. *Eur J Gastroenterol Hepatol* 1996; 8: 657-61.
4. Cantarini D, Carli G, Dalla Libera M, Gullini S. Aspetti clinici del morbo di Crohn. In: Gullini S, Cantarini D. Rettocolite ulcerosa e morbo di Crohn. Edizioni Ferraria Libro, Ferrara 1993, 37-45.
5. Bradley PJ, Ferlito A, Devaney KO, Rinaldo A. Crohn's disease manifesting in the head and neck. *Acta Otolaryngol* 2004; 124: 237-41.
6. Pellicano R, Bonardi R, Smedile A, et al. Poliambulatori specialistici di gastroenterologia: l'esperienza 2003-2006 dell'Ospedale Molinette di Torino. *Minerva Medica* 2007; 98: 19-23.
7. Astegiano M, Bresso F, Demarchi B, et al. Organisation of work teams at the gastroentero-hepatology outpatients clinic: the management of inflammatory bowel diseases. *Minerva Gastroenterol Dietol* 2002; 48: 1-5.

Accepted: April 27th 2011
Correspondence: Dr. Rinaldo Pellicano
Department of Gastro-Hepatology
Via Chiabrera 34, III floor
10100 Turin, Italy
E-mail: rinaldo_pellican@hotmail.com