CASE REPORT

A RARE CASE OF ISOLATED OESOPHAGEAL CROHN’S DISEASE

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Abstract. Crohn’s disease (CD) may affect any segment of the gastrointestinal tract. Although rare, its oesophageal manifestation may be seen as the first sign of this disease. We present the case of a 27 years old female who is admitted with heartburn and dysphagia for 4 years, being misdiagnosed as reflux esophagitis. Endoscopy performed in our unit pointed out erythema, segmental ulcers separated by normal mucosa, “cobble stone” aspect. The histological findings were ulcers and a chronic inflammatory mucosal infiltrate. The patient was unresponsive to PPI(Proton Pump Inhibitor), but a rapid improvement was seen after corticosteroid administration. Differential diagnosis and treatment opportunities are presented below.

Keywords: esophageal Crohn’s disease, esophageal ulcers, corticosteroids

Introduction

Crohn’s disease is a chronic inflammatory disease of unknown aetiology characterized by a chronic, granulomatous, segmental transmural inflammation that may affect any part of the gastrointestinal tract. The ileum, the colon, the rectum, and the perianal region are the most frequent locations of disease involvement. Esophageal Crohn’s disease is rare, with an adult prevalence of 0.2% to 3% in patients with coexisting ileocolonic disease. Very few cases of isolated esophageal involvement are reported.[1,2] An accurate diagnosis and treatment is often made rather late in its course, due to the unusual presentation of isolated esophageal CD. Here, we report a case of isolated esophageal CD, misdiagnosed as reflux esophagitis, unresponsive to PPI, with a favorable clinical outcome after oral corticosteroids.

Patient, methods and result

A 27 years old patient presents with dysphagia and heartburn for 4 years persistent despite PPI treatment. She didn’t manifest any modification in the stool pattern. She was non smoker and denied alcohol consumption.

She was examined in the past by three upper endoscopy exams which were interpreted as reflux esophagitis. Blood tests revealed slight inflammatory syndrome (CRP-10 mg/dl) and no other modification.

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Fig.1. (a, b, c)-endoscopy images

We repeated an upper gastro-intestinal endoscopy which showed throughout the entire length of the oesophagus, oedema, erythema, segmental ulcers with a cobble stone aspect. Biopsies were sampled (Figure 1). Colonoscopy was normal on the entire colon, also terminal ileum was without lesions. Abdominal CT scan was also normal (for suspected lesions of small bowel).
Oesophageal mucosa biopsy fragments collected from the oesophageal mucosa showed active esophagitis, erosions, non granulomatous with reactive cellular changes and regional aspects of basal layer squamous epithelial hyperplasia, intraepithelial frequent PMN neutrophils; in the lamina propria of the esophageal mucosa areas with granulation tissue, marked polymorphous inflammatory infiltrate and lymphoid follicle with germinal center. No evidence of granuloma, cytopathic changes suggestive of a viral or neoplastic context on the fragments examined. The PAS stain highlights no pseudohyphae. Therefore, histological features suggested active Crohn's disease.

She was treated by oral corticosteroids with significant alleviation of dysphagia and disappearance of heartburn.

According to the ECCO (European Crohn's and Colitis Organisation) oesophageal or gastroduodenal Crohn's disease may best be treated with a proton pump inhibitor, if necessary together with systemic corticosteroids and thiopurines or methotrexate. Anti-TNF therapy is an alternative for severe or refractory disease. Dilatation or surgery is appropriate for obstructive symptoms [3].

Hence we have started thiopurine therapy with Azathioprine, without any adverse reaction encountered by the patient. The endoscopic mucosal remission will be evaluated in at least 3 months, the necessary period of Azathioprine action. If she fails to respond to Azathioprine, anti TNF agents will be introduced.

**Discussions**

Almost all the esophageal CD reported in literature has coexisted with CD at other sites, such as the ileum, rectus and colorectum [4]. Here, we report the case of a patient with isolated esophageal CD.

In general, CD of the oesophagus is not difficult to diagnose in cases in which other segments of the
digestive tract are also involved, or in patients with a prior history of CD.

The histopathological description of oesophageal Crohn's disease is generally nonspecific, represented by oesophageal mucosal ulceration, exulcerations, reactive changes of oesophageal squamous epithelium, increased number of intraepithelial lymphocytes and polymorphonuclears, fibrosis and inflammatory lymphocytic infiltration in lamina propria.

The presence of noncaseating granulomas associated to esophagitis in a patient with known Crohn's disease, affecting other segments of gastrointestinal tract is a clear sign of oesophageal Crohn's disease.

The presence of granulomas associated to esophagitis, without any other lesions of gastrointestinal tract requires differential diagnosis with infectious mycobacterium esophagitis or sarcoidoisis. In determining infectious aetiology of esophageal lesions is useful to identify viral cytopathic changes characteristic of herpes simplex virus and cytomegalovirus; also PAS staining in fungal esophagitis may reveal pseudohyphae. Histopathological aspect of active esophagitis, erosive, nongranulomatous may be also present in gastro-oesophageal reflux disease, ingestion of corrosive agents, drug-induced esophagitis, in motility disorders (achalasia), collagen diseases.

Lymphocytic esophagitis associated particularly with Crohn's disease in paediatric pathology involves an increased number of intraepithelial lymphocytes, but without the presence of intraepithelial polymorphonuclear cell [5]. Eosinophilic esophagitis is diagnosed histologically by an increased number of eosinophils in the esophageal squamous epithelium [6]. Treatment of proximal CD depends on the severity of the disease. The ECCO consensus underscores the lack of controlled studies to demonstrate the effectiveness of drug therapy and proposes the addition of a proton pump inhibitor in the conventional treatment for distal disease or the use of systemic corticosteroids and thiopurines or methotrexate [3]. Antisecretory drugs (histamine receptor antagonists; HRAs and proton-pump inhibitors; PPIs) may relieve pain, but their efficacy in mucosal healing has not been proven. Corticosteroids as a systemic therapy are the best treatment of choice, while no data exist on the therapeutic effect of budesonide formulations. Immunosuppressive therapy with azathioprine, 6-mercaptopurine or methotrexate is an excellent steroid-sparing option for long-term maintenance of the disease. The ECCO consensus which suggested that clinicians should have a lower threshold for starting anti-TNF therapy than for disease elsewhere, given the poor prognosis of Crohn's disease with proximal involvement [7, 8].

Conclusion

Isolated Crohn's disease is very rare and may be underdiagnosed. The endoscopic aspect is highly suggestive and it is based on the endoscopist's experience of recognizing the lesions. As a result, our patient had three endoscopic exams prior to the presentation, misdiagnosed as reflex esophagitis. Also the histological exam is challenging and requires differential diagnosis with other entities. Rapid clinical improvement after corticosteroids highlights the accuracy of diagnosis.

References

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