Letters to the Editor

Multiple perforation of small-intestine diverticula in a patient with Ehlers-Danlos syndrome

**Key words:** Ehlers-Danlos. Diverticulum. Duodenum.

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Dear Editor,

Ehlers-Danlos syndrome represents a group of hereditary connective tissue disorders characterized by ligamentous hyperlaxity, fragile skin and joint hypermobility. Gastrointestinal complications in this syndrome are less well known (1).

**Case report**

We report the case of a 42-year-old male with this condition who presented with acute abdominal over a 12 hour period after taking anti-inflammatory drugs. A CT scan revealed a right-sided retro-hydro-pneumoperitoneum secondary to duodenal diverticular perforation extending to the falciform ligament. An exploratory laparotomy was performed, and biliary peritonitis from two perforated duodenal diverticula were found: one with three perforations, the other with two. Pathological analysis of the specimen found that it contained duodenal segments with no muscularis propria and parietal necrosis, which confirmed the diagnosis (Fig. 1). The patient had a favorable outcome following the procedure, and was discharged within a few days.

**Discussion**

Most common gastrointestinal manifestations in patients with Ehlers-Danlos syndrome include abdominal pain, nausea and vomiting (2), and gastrointestinal bleeding and intestinal perforation may be seen in severe cases (3). A higher incidence of colonic diverticulosis has also been described for this condition, a fact thought to result from a lack of distension at the colonic wall as a consequence of collagen impairment, but the specific mechanism still remains unknown (2,4). In this patient, however, diverticula were predominantly located in high numbers within the small bowel. Perforations may be spontaneous or brought about by some external factor (anti-inflammatory drugs, endoscopic instrumentation), and their multiplicity even in one diverticulum or apparently healthy tissue must be highlighted (5).

Therefore, excluding a diagnosis of intestinal diverticulosis and its related complications is crucial for patients diagnosed with Ehlers-Danlos syndrome presenting with abdominal pain.

Raúl Honrubia-López1, Aurora Burgos-García1 and Elena Palacios-Lázaro2

Fig. 1. A. Abdominal CT showed a duodenal diverticulum (arrow) and retro-hydro-pneumoperitoneum. B. Duodenal segment with no muscularis propria that displays wall necrosis and severe acute peritonitis.
Departments of Digestive Diseases, and 2Pathology. Hospital Universitario La Paz. Madrid, Spain

References


