Clinical Case Report

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Small bowel capsule endoscopy revealing neuromuscular and vascular hamartoma of the jejunum

A case report

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Abstract

Rationale: Digestive hemorrhage is a life-threatening and represents for both clinicians and patient a challenger problematic condition with the urgencies to discover the origin for correct the cause and safe the life of patient.

Patient concerns: We report the case of a 58 -year-old man with extremely rare hamartomatous neurovascular lesion. Following recurrent episode of intestinal hemorrhage the patient underwent small bowel capsule endoscopy.

Diagnoses: Diagnosed with small intestine neoplasia.

Interventions: The patient underwent curative small bowel resection. Histologic diagnosis was neuromuscular and vascular hamartoma (NMVH). In the small intestine, neoplastic lesions are very rare (2%) and mostly malformative while the more frequent cause of cryptic digestive hemorrhage remains angiodysplasia (50%) . The preexisting NMVH was exacerbated by the use of non-steroidal anti-inflammatory drugs, causing hemorrhage due to diffuse ulceration.

Outcomes: The patient stay healthy after treatment.

Lessons: This is an hemorrhagic lesion with macroscopic "neoplastic" patterns due to abnormal mixing of normal indigenous tissue components. It poses a diagnostic challenge for clinicians and pathologists, but diagnosis is facilitated by capsule endoscopy and surgical treatment should provide definitive resolution.

Abbreviations: GI = gastrointestinal, NMVH = neuromuscular and vascular hamartoma, NSAID = nonsteroidal antiinflammatory drug.

Keywords: capsule endoscopy, digestive hemorrhage, neuromuscular and vascular hamartoma (NMVH)

1. Introduction

Digestive hemorrhage is an emergency life-threatening, challenging condition, and it is essential to rapidly discover the origin to correct the cause and safeguard the patients' life. Gastrointestinal (GI) bleeding is one of the most serious GI tract conditions, with a mortality rate of 10%.[1] Capsule endoscopy is most commonly performed for obscure GI bleeding, but may also be used in the evaluation and surveillance of patients with hereditary polyposis syndromes, small bowel damage induced by nonsteroidal antiinflammatory drugs (NSAIDs), the diagnosis and follow-up of Crohn disease, and the suspicion of a small bowel tumor and celiac disease. Occult GI hemorrhage, defined as bleeding that remains unexplained after a bidirectional negative endoscopic evaluation of the GI tract, accounts for approximately 5% of all GI bleeding.[2] For small bowel tumors, capsule endoscopy evaluation is not considered due to the elevated risk of capsule retention. The main cause of digestive hemorrhage of the upper GI tract is peptic ulcer, accounting for 28% to 59% of cases.[3] The most frequent cause of occult hemorrhage is angiodysplastic lesions, present in 50% of cases; in particular sites like the small intestine, it is difficult to diagnose. Neuromuscular and vascular hamartoma (NMVH) is a rare hemorrhagic lesion of the intestine, with only 23 cases reported in the English literature since its initial description by Fernando and McGovern in 1982.[4–7] The clinical presentations of NMVH patients are usually nonspecific, including abdominal pain, obstructive symptoms, occult GI bleeding, and iron deficiency anemia. The lesion consists in a disorganized benign mass of indigenous cells resulting in an aberrant submucosal proliferation of muscular, neural and vascular elements (hemangiomatous vessels), affecting a variable length of bowel, and causing stenosis. Surgical resection is curative,[8] and there have been no documented recurrences of NMVH owing to the malformative, nontumoral nature of the disease.

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Compliance with ethical standards.

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1.1. Consent statement

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

2. Case presentation

We report the case of a 58-year-old man complaining of recurrent diffuse abdominal pain, with a positive fecal occult blood test, iron deficiency with mild anemia, and presenting vomiting and an episode of high fever associated with mildly increased ESR and PCR. *Anisakis* was suspected due to his predilection for raw fish, but ample parasitological analyses were negative. The patient had a medical history of NSAID use for a herniated disc. Biological markers, gastro-duodenal, and colon endoscopy excluded IBD and other more frequent hemorrhagic causes. Following a 2nd episode of intestinal hemorrhage the patient underwent small bowel capsule endoscopy (SBCE–PillCam SB3 system): at 2 hours 38 minutes from the beginning of the recording, the examination demonstrated an ulcerative lesion on the jejunum, associated to an evident neoformation protruding into the lumen (Fig. 1A). Macroscopically diagnosed as an undefined tumor of the small intestine, surgery was scheduled. After excluding Crohn disease in the absence of the classical clinical-pathological and serological features, the hemorrhagic lesion was surgically treated by curative resection. This is also therapeutically effective in small bowel angiodysplasia, that is more frequently observed. Macroscopically, the 6.5 cm segment of small bowel mucosa appeared as a granular, mucoid bump with an ulcerated neighboring area. Microscopically, the mucosa showed focal intramucosal hemorrhage; in the submucosa under the large ulcerated area the vascular hyperplastic component exhibited wall thickening with endoluminal proliferation and obliteration (Fig. 1B). The elevated area showed a true arteriovenous malformation with a prominent neuroganglionic component: deformed vessels and distorted dilated thin-walled venules mostly lined only by endothelium (Fig. 1C) and frequently a small amount of smooth muscle, crossing the muscolaris mucosae. The submucosa was strongly expanded by disorganized bundles of smooth muscle in continuity with the muscularis mucosae and scattered abnormal proliferations of neurogangliar structures intermingled with ectasic thick-walled vascular channels (Fig. 1D). The final histopathology diagnosis was NMVH as described by Fernando and McGovern.[4] The patient's current state is fine and does not need follow-up for 2 reasons: the kind of small bowel examination with the capsule endoscopy excluded the presence of the other mass protruding in the lumen; in literature, multiple MNVH lesions have not been reported.

3. Discussion

The pathological findings were similar to those described for NMVH, a rare lesion of the intestine.[4–7] The hamartomatous nature of this disorder is questioned by many authors because similar features may be a part of the histological spectrum of Crohn disease, NSAIDs-associated small intestinal diaphragm disease, and ischemic and radiation enteritis.[9] Shepherd and Jass[10] suggested that NMVH may represent "an unusual histologic consequence of inflammatory bowel disease, predominantly Crohn disease." Moreover, neuromatous dysplasia and ganglionitis is well known to GI pathologists as additional patterns of Crohn disease, but in the absence of other classical

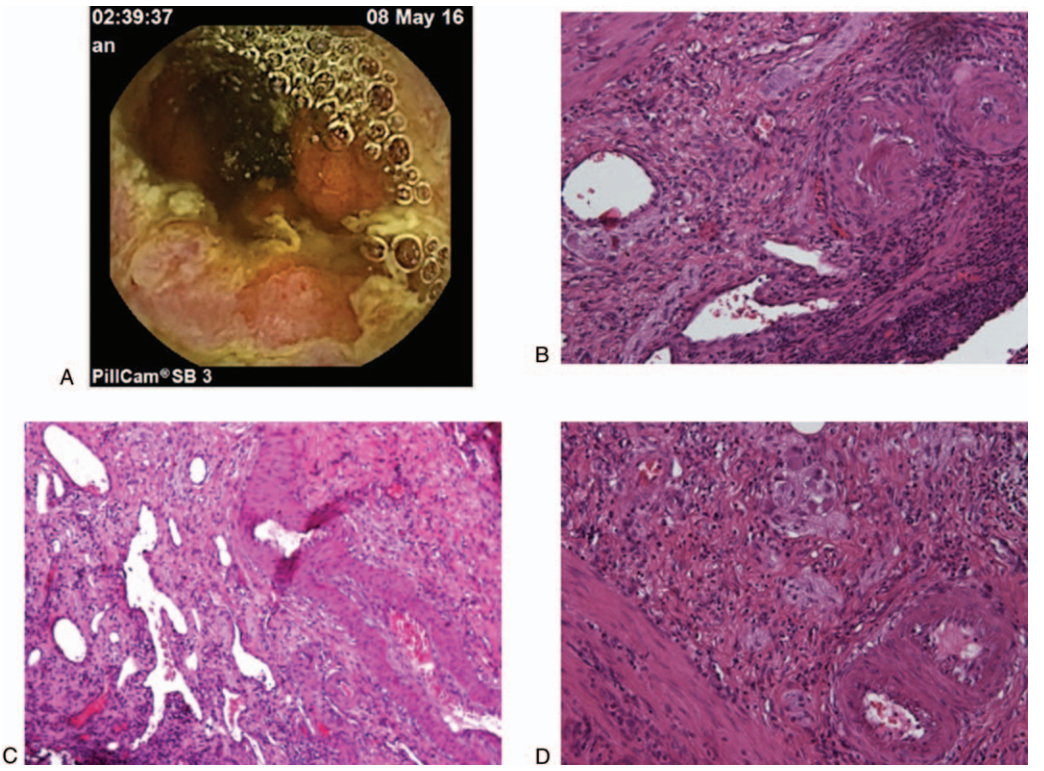

Figure 1. (A) Ulcerative lesion on the jejunum, associated to an evident neoformation protruding into the lumen. (B) Vessel wall thickening with endoluminal proliferation and obliteration, (C) deformed vessels and distorted dilated venules, and (D) neurogangliar structures intermingled with ectasic thick-walled vascular channels.

features related to Crohn, the final diagnosis was NMVH. At first, the NMVH mass-forming lesion may seem to be a neoplastic lesion. In the small intestine, neoplastic lesions are very rare (2%) and mostly malformative.[11] However, the most frequent cause of cryptic hemorrhage remains angiodysplastic lesions, that are particularly difficult to diagnose in the small intestine. Angiodysplasia may account for approximately 6% of cases of lower GI bleeding, while small bowel angiodysplasias may account for 50% of obscure GI bleeding.[12] In a recent retrospective colonoscopic analysis, it was shown that 12.1% of 642 patients without symptoms of irritable bowel syndrome, and 11.9% of those with irritable bowel syndrome had colonic angiodysplasia.[13] Certainly, in this case the preexisting NMVH was exacerbated by the use of NSAIDs, causing hemorrhage due to diffuse ulceration of the mucosa covering hamartomatous lesion. NMVH remains a controversial entity, as most authors consider it a remnant of chronic "burnt-out" Crohn[9] caused by chronic use of NSAIDs.[14,15]

In conclusion, this is the first case in literature of NMVH discovered thanks to capsule endoscopy, and including the related images. NMVH is an extremely rare lesion with macroscopic "neoplastic" patterns and a prevalently severe hemorrhagic clinical presentation due to abnormal mixing of normal indigenous tissue components. It poses a diagnostic challenge for clinicians and pathologists, but the diagnosis is facilitated by capsule endoscopy and surgical treatment should provide definitive resolution.

Author contributions

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