FIRST CASE OF SERRATED LESION OF THE DUODENAL PAPILLA IN A PATIENT WITH SERRATED COLONIC POLYPOSIS

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Conflicts of interest: Marine Camus is a consultant for Boston Scientific and Cook Medical. Xavier Dray is cofounder and shareholder of Augmented Endoscopy and has acted as a consultant for Alfasigma, Bouchara Recordati, Boston Scientific, Fujifilm, Medtronic, and Pentax. The other authors have no conflicts of interest to disclose.
This work was not sponsored by gifts or fellowships.

Keywords: Serrated polyposis, duodenal serrated lesion, screening endoscopy procedures

ABSTRACT

A serrated polyposis syndrome was diagnosed in a 26-year-old female presenting with gastrointestinal symptoms. Screening for other lesions of the gastrointestinal tract showed a serpiginous looking papilla, described as possibly dysplastic. Histological analysis of biopsies showed a serrated lesion. This case describes the first known association between a duodenal serrated lesion and serrated polyposis syndrome. Upper GI screening is probably of little interest in this setting. In patients with upper GI serrated lesions, we recommend screening colonoscopy.

INTRODUCTION

Serrated polyposis syndrome (SPS) is a colorectal polyp syndrome characterized by a combination of large and/or numerous serrated lesions, and is associated with an increased risk of colorectal cancer (CRC). Duodenal polyps have been described in the setting of colorectal polyp syndromes, but not in patients with SPS. We describe in this clinical case the first association between SPS and a duodenal serrated lesion.

CASE PRESENTATION

A 26-year-old female patient was initially referred for alternating diarrhea and constipation as well as intermittent abdominal pain. Past medical history was unremarkable. Physical examination was unremarkable. A colonoscopy was performed and showed multiple flat and sessile polyps, located throughout the different segments of the colon, ranging from 5 to 20 mm in diameter. The macroscopic aspect was compatible with serrated lesions as they presented with an irregular cloud-like surface, with black pits on some of them, and because a disruption of the vascular pattern of the surrounding colonic mucosa was noted (**Figure 1A**). Histological assessment confirmed these were sessile serrated lesions (SSL) with low-grade dysplasia (**Figure 1B to 1G**), establishing the diagnosis of SPS. After a multidisciplinary staff, resection of all the polyps located in the rectum and sigmoid colon was decided, before eventual subtotal colectomy. A complete endoscopic examination was performed in order to screen for other lesions of the gastrointestinal (GI) tract. Upper GI endoscopy and side-viewing duodenoscopy unexpectedly showed a serpiginous looking major papilla measuring 20 mm in length, described as possibly dysplastic (**Figure 2A**). Histological analysis of biopsies showed a serrated non-dysplastic lesion with the same immunophenotype as the colonic serrated adenomas/polyps (**Figure 2B to 2F**). Colonoscopy allowed resection of multiple polyps in the rectum and sigmoid colon. A subtotal colectomy with ileosigmoid anastomosis was

subsequently performed. Analysis of the specimen confirmed multiple SSLs (about a hundred) and no cancerous lesion. The patient has undergone annual surveillance since. The last surveillance endoscopy was performed less than a year ago, with resection of three non-dysplastic SSLs of the sigmoid colon and a similar endoscopic and histologic aspect of the major papilla. After they were all contacted, both parents and one of the five siblings of the patient underwent screening endoscopies (EGD, lateral duodenoscopy, and colonoscopy). All three family members had normal appearing major papillas. However, the father and one brother were diagnosed with SPS. A genetic testing is currently underway.

DISCUSSION

SPS has been defined by the World Health Organization (WHO) in 2010 as the presence of (1) ≥ 5 histologically diagnosed serrated lesions proximal to the sigmoid colon, of which ≥ 2 are larger than ≥ 10 mm; or (2) any number of serrated polyps proximal to the sigmoid colon in an individual who has a first-degree relative with SPS; or (3) ≥ 20 serrated polyps distributed throughout the colon [1]. The true prevalence of SPS is unknown but it is likely underestimated, at 0.9% by recent studies [2]. The prevalence of CRC in this setting ranges between 15% and 35% [3,4]. Given the risk of malignant progression, including during surveillance, endoscopic surveillance is recommended for all patients via colonoscopy every 1 to 2 years [5,6].

Studies have evaluated whether SPS could harbor a risk of extracolonic polyposis or cancers. In a study published in 2013 including 44 patients with SPS, 22 patients had an esogastroduodenoscopy (EGD), with no lesion found [7]. In a cohort including 51 patients with SPS, published by Jasperson et al. in 2013, 30 patients had underwent an EGD, with no lesion found either [8]. There does not seem to be an extracolonic cancer risk either in patients with SPS [9,10]. The British guidelines state that '*upper GI surveillance for polyposis or extraluminal*

surveillance for non-GI cancers is not necessary in patients with SPS where other genetic causes have been excluded' [5].

Regarding serrated lesions of the duodenum, data are scarce. The first case of duodenal SSL associated with a hereditary CRC syndrome (familial adenomatous polyposis) was published by Rubio et al. in 2004[11]. Several cases of duodenal serrated lesions have been published since, mostly of hyperplastic polyps [12–14]. A study by Cao et al. evaluated an endoscopy database of 98,746 patients. Serrated lesions of the upper GI tract where found in 21 patients (0.02%) and 3 lesions were found in the duodenum only [15]. Overall, serrated lesions of the duodenum are rare, which accounts for the fact that their natural history and prognosis are unknown and the appropriate management unclear.

To our knowledge, the association between SPS and duodenal serrated lesions has not been reported so far. In the previously mentioned study by Cao et al., colonoscopy data were available for 18 of the 21 patients with upper GI serrated lesions and none had features of SPS although a higher colorectal adenoma detection rate was observed compared to patients with no upper GI serrated lesions [15]. We describe the first association between SPS and a duodenal serrated lesion (SSL). This case suggests that although extremely rare, patients with SPS can harbor upper GI lesions. However, based on the description of one case, we do not believe that EGD should be performed in patients with SPS in a systematic manner. However, there may be a subgroup of SPS patients in whom this screening might be of interest. Further studies are warranted to clarify the clinical significance of these lesions, such as genetic analysis to identify pathways associated with SPS. Conversely, based on the results of the study by Cao et al., we would recommend that patients with serrated lesions of the upper GI tract should undergo colonoscopy, as well as their first-degree relatives.

In conclusion, we report the first case of a serrated lesion in the duodenum associated with SPS. Upper GI screening is probably of little interest in this setting. However, in patients with serrated lesions of the upper GI tract, we would recommend screening colonoscopy.

FIGURES

Figure 1: Serrated lesion of the colon

Endoscopic image in near focus and virtual chromoendoscopy (Narrow Band Imaging) showing a slightly elevated polyp located in the ascending colon (Paris IIa), with a maximal diameter of 10 mm, an irregular cloud-like surface, with a few black pits (A). Histological assessment showed a distortion of the normal architecture (D, HE x 50) with dilated L-shaped or inverted T-shaped crypts and serrated glands (B, HE x 100). Serrated dysplastic areas were present (C, HE x 200). Immunohistochemical studies showed immunoreactivity for Annexin A10 (E, x 100), MUC2 (F, x 100) and MUC5AC (G, x 100). MLH1 expression was not lost (not shown).

Figure 2: Serrated lesion of the duodenal papilla

Endoscopic image in white light (A1) and virtual chromoendoscopy (Narrow Band Imaging) (A2) showing a serpiginous appearing papilla. Histological assessment showed a polypoid lesion (C, x 50) with slightly serrated epithelium without dysplasia (B, x 100). As the colonic serrated adenomas/polyps, immunoreactivity for Annexin A10 (E, x 100), MUC2 (F, x 100) and MUC5AC (G, x 100) was observed, and MLH1 expression was not lost (not shown).

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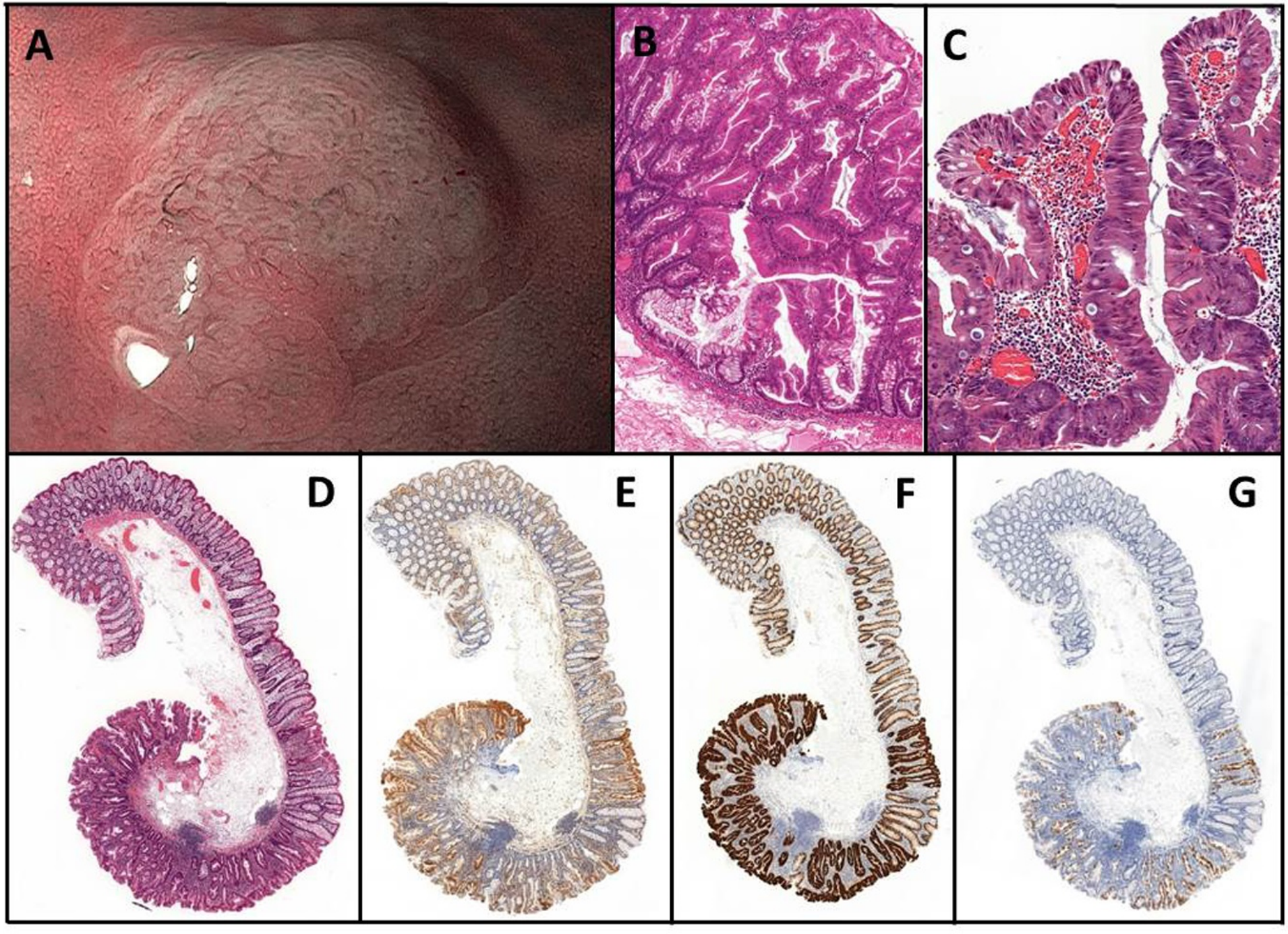
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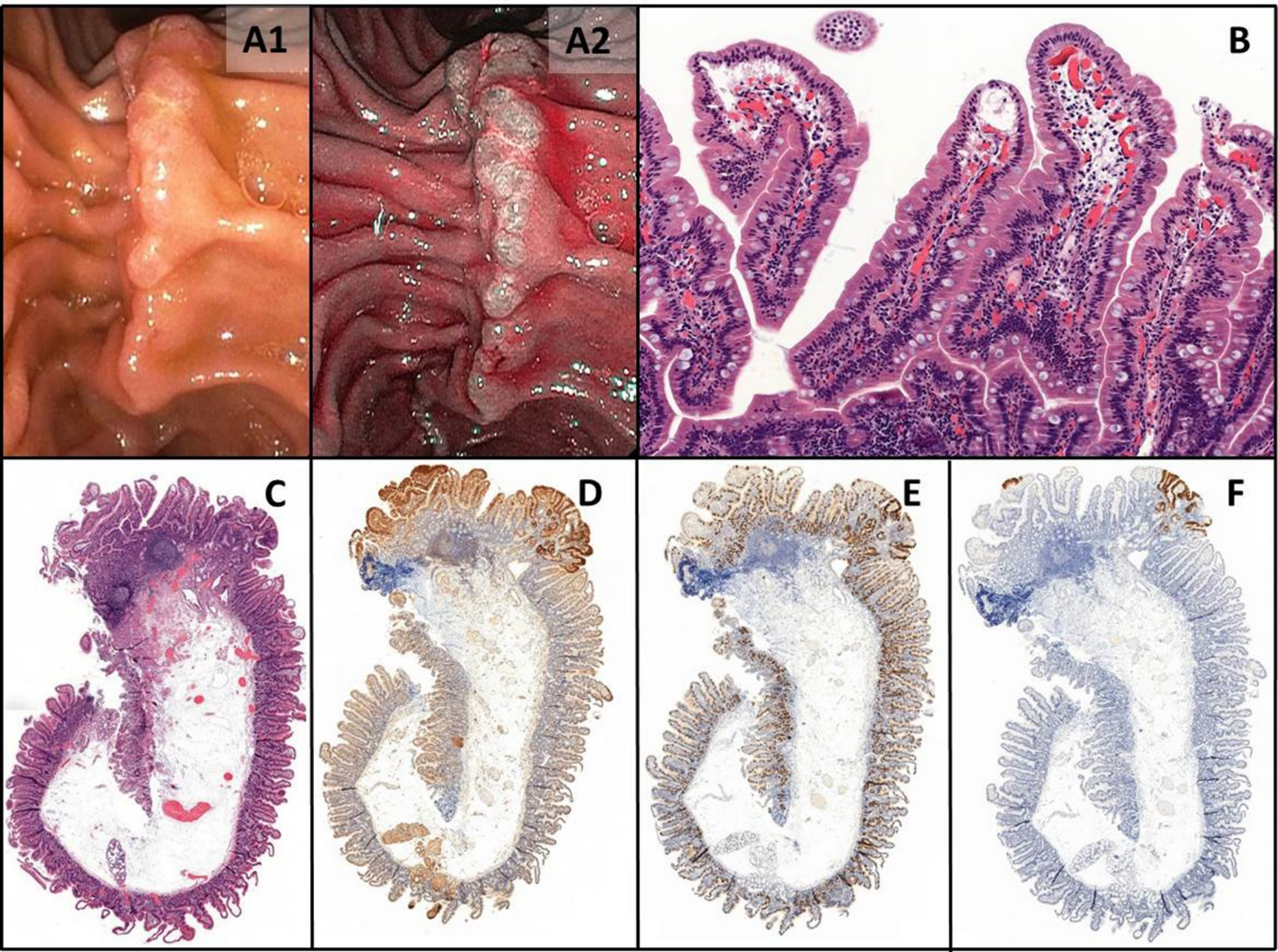
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