

A laparoscopic right hemicolectomy was performed, without finding any extraluminal or exophytic abnormalities, and the patient was discharged home six days later.

The pathology report was of a tumour-like lesion in the appendiceal *ostium*, compatible with endometriosis at the base of the implantation of the appendix, ruling out malignant disease. The patient was asymptomatic at subsequent follow-ups.

Endometriosis is the presence of endometrial glands and stroma outside the uterine cavity. There is great debate over the pathogenesis. Three theories are postulated: coelomic metaplasia, retrograde menstruation and ectopic endometrium resulting from diffusion through lymphatic and blood vessels.<sup>3</sup>

It can be classified as pelvic or extra-pelvic, with pelvic endometriosis being limited to the fallopian tubes, ovaries and nearby pelvic peritoneum, while extrapelvic endometriosis includes intestinal endometriosis in the urinary system, skin, lungs, liver and even the heart.<sup>4</sup>

Endometriosis of the gastrointestinal tract accounts for 3% to 15% of all cases.<sup>5</sup> It is most often located in the rectosigmoid region, sometimes causing non-specific symptoms, including rectal bleeding, haematochezia, pain and obstruction, which can lead to the patient being screened for cancer.

The involvement of the appendix in endometriosis is uncommon, representing 2–3% of gastrointestinal endometriosis and 1% of endometriosis overall,<sup>5</sup> many cases being diagnosed after an appendectomy for suspected appendicitis. In the case we describe here, due to the patient's gastrointestinal history, imaging tests and non-specific pathology results, we were directed towards possible appendix cancer, probably a carcinoid tumour, as that is the most common type.<sup>1</sup> We decided to perform a right laparoscopic hemicolectomy and only learned the actual diagnosis, endometriosis with caecal implantation, when we received the pathology results.

The literature describes endometriosis involving the appendix mainly at the level of the body and tip, affecting the serous or submucosal layer. Endoluminal involvement<sup>5</sup> and location in the appendiceal *ostium* is very rare. The similarity in imaging tests with a carcinoid tumour, along with its location, and the lack of a pathognomonic image for diagnosis, mean it is unlikely to be suspected prior to

surgery. Although in our case, the endometriosis was in the appendiceal *ostium*, it can present as acute appendicitis, intestinal perforation, intussusception or as a lower gastrointestinal bleeding, and it can mimic cancerous processes such as appendiceal mucocoeles or carcinoid tumours. We draw attention to the back and forth between the diagnostic options of ileocaecal and appendiceal lesions to endometriosis. Although it represents a diagnostic challenge pre-operatively, it can help avoid radical or unnecessary surgery.

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## Immune thrombocytopenic purpura associated with inactive ulcerative colitis in chronic treatment with adalimumab<sup>☆</sup>



### Púrpura trombocitopénica inmune asociada a colitis ulcerosa inactiva en tratamiento crónico con adalimumab

We read with great interest the letter published in your journal from Brunet et al.<sup>1</sup> As the authors comment, immune

thrombocytopenic purpura (ITP) is a rare haematological extra-intestinal manifestation in inflammatory bowel disease (IBD). We present the case of a female patient with a history of ulcerative colitis for many years who developed ITP despite long-term treatment with adalimumab and no gastrointestinal activity. This seems to be a particularly exceptional case and makes us reflect on the different aetiopathogenic mechanisms involved in the condition.

This was a 54-year-old woman diagnosed with left-sided ulcerative colitis in 2006. Over the course of her disease, she had been treated with mesalazine and azathioprine from 2012, with an admission in 2013 due to a severe flare-up, with no response to infliximab, but which did resolve with adalimumab at the usual dose, being increased to 40 mg weekly in autumn 2014 due to clinical activity. In December 2016, the patient was asymptomatic with complete blood count normal. In January 2017, she was admitted

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with rectal bleeding without diarrhoea but with anaemia, with haemoglobin levels of 7.7 g/dl and platelets 4000/ $\mu$ l. Her spleen appeared normal on ultrasound. Haematology completed investigations of acute thrombocytopenia with a final diagnosis of ITP and she started treatment with platelet transfusion, gamma globulin 1 g/kg/day for two days and prednisone 1 mg/kg/day, in tapering regimen for two months, during which time the patient's platelet levels returned to normal. The test for *Helicobacter pylori* was negative, as were the viral and autoimmunity determinations. A full colonoscopy was performed, showing no signs of activity, but rectal angiodysplasia, which was treated with argon. Azathioprine and adalimumab were discontinued. However, due to clinical-endoscopic flare-up when the patient stopped prednisone, it was decided to reintroduce them once again. Since then she has remained asymptomatic and with normal platelet count.

The incidence of ITP in patients already diagnosed with IBD is 12.5/100,000 population/year, apparently in relation to the severity of the gastrointestinal disease. The prevailing aetiological hypothesis is an increase in the permeability of the mucosa due to inflammation, with the passage of bacterial antigens into the circulation giving rise to a secondary, systemic, Th1 humoral response.<sup>1</sup> It has been suggested that there may be antigenic mimicry between some platelet surface proteins and bacterial glycoproteins, resulting in a cross-reactive immune response. Another mechanism could be the sequestration and subsequent direct destruction of platelets in the increased microvascularisation of the colon, which could explain the lack of antibodies, the resistance to corticosteroids and the resolving of the problem with colectomy found in some cases.<sup>2</sup> These two theories would explain why ITP occurs more frequently during a flare-up of IBD. However, in our patient, the ITP developed suddenly and while her IBD was inactive.

This situation led us to consider a possible relationship between the onset of her ITP and her baseline medication, despite the lack of recent changes. Although the azathioprine and mesalazine she was taking can cause thrombocytopenia due to bone-marrow toxicity, it was classified as peripheral by Haematology. Severe adalimumab-induced thrombocytopenia is recognised as anecdotal and mostly in patients taking it for disorders other than IBD<sup>3</sup>; an incidence of 0.1 % for events with < 100,000 platelets is reported for adalimumab.<sup>4</sup> The onset of thrombocytopenia after being on adalimumab for over two years and its non-recurrence in controlled reintroduction would go against the likelihood of drug-related aetiology in our patient.

Other authors support the existence of a genetic relationship between IBD and ITP through the major histocompatibility complex, as the incidence of ITP in patients with IBD is higher than in the general population and is

accepted as not being fortuitous. Polymorphisms have been described in genes encoding immunity which could be shared between ITP and IBD.<sup>5</sup>

In conclusion, ITP is a very rare condition which can be associated with IBD. We have presented a case of such an association with highly unusual characteristics, including the fact that it occurred despite a lack of gastrointestinal activity and the patient being on treatment with adalimumab, and the fact that it did not recur after reintroducing adalimumab. These circumstances expose the lack of accurate data on the ultimate pathogenic mechanism by which both disorders associate and interrelate.

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