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Introduction

Inflammatory bowel disease (IBD) is associated with a higher risk of developing gastrointestinal neoplasms, among them adenocarcinoma is the most common subtype. Although less frequent, IBD patients also present higher incidence of neuroendocrine tumors (NETs) compared to the general population[1].

Case report

We describe the case of a 62-year-old man with ileal Crohn's disease (CD) treated with azathioprine since 2018 who continued with inflammatory activity with no further follow-up.

He consulted for abdominal pain and lower limbs edema. Laboratory tests revealed a moderate hypokalaemia (2.5 mEq/L) and a mild hyperbilirubinemia (1.33 mg/dl). Abdominal ultrasound showed multiple liver lesions suggestive of metastasis and signs of ileitis, with two abdominal collections adjacent to the ileum. These findings were confirmed by computerized tomography.

Corticosteroids and antibiotics were started with an improvement of abdominal symptoms and acute phase reactants. However, the patient suffered from hypernatremia, hypokalaemia, hyperglycaemia, and lower limbs edema.

Abdominal magnetic resonance imaging scan showed an asymmetric thickening of the ileum, that was suspected as the primary tumor, and the same metastatic liver lesions. Ileocolonoscopy showed typical signs of CD. Histopathological exam of liver biopsy showed infiltration of large-cell NET.

Blood adrenocorticotropic hormone (ACTH) levels were >1500 pg/mL, so an ectopic Cushing syndrome (CS) was suspected and treatment with ketoconazole was started.

An Octreoscan was requested but the patient had a destabilizing gastrointestinal bleeding that required multiple blood transfusions. The ileal tumor was suspected as the origin of the bleeding, and an urgent surgical resection was performed. The histopathological study of the resection specimen confirmed a grade 3 NET (well-

differentiated morphology with Ki-67 of 80%). After multiple postoperative complications, the patient died.

Discussion

NETs are a heterogeneous group of neoplasms whose most frequent location is the gastrointestinal tract and pancreas, followed by lungs[2]. Some differentiated NETs can rapidly proliferate and form distant metastasis. Given their low incidence, high heterogeneity and low index of suspicion, NETs remain a late-diagnosed entity in many cases[3].

Paraneoplastic Cushing syndrome (PCS) is caused by ectopic tumor production of ACTH or its releasing hormone (CRH). It accounts for 10% of CS cases. However, PCS is extremely rare in gastrointestinal NETs, but more common in pulmonary NETs and small-cell lung cancer[3].

Metastatic jejunum and ileum NETs have a low survival rate, but resection of the primary tumor provides significant benefits. Nevertheless, PCS is associated with worse outcomes due to a higher complication rate[4,5].

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Figures

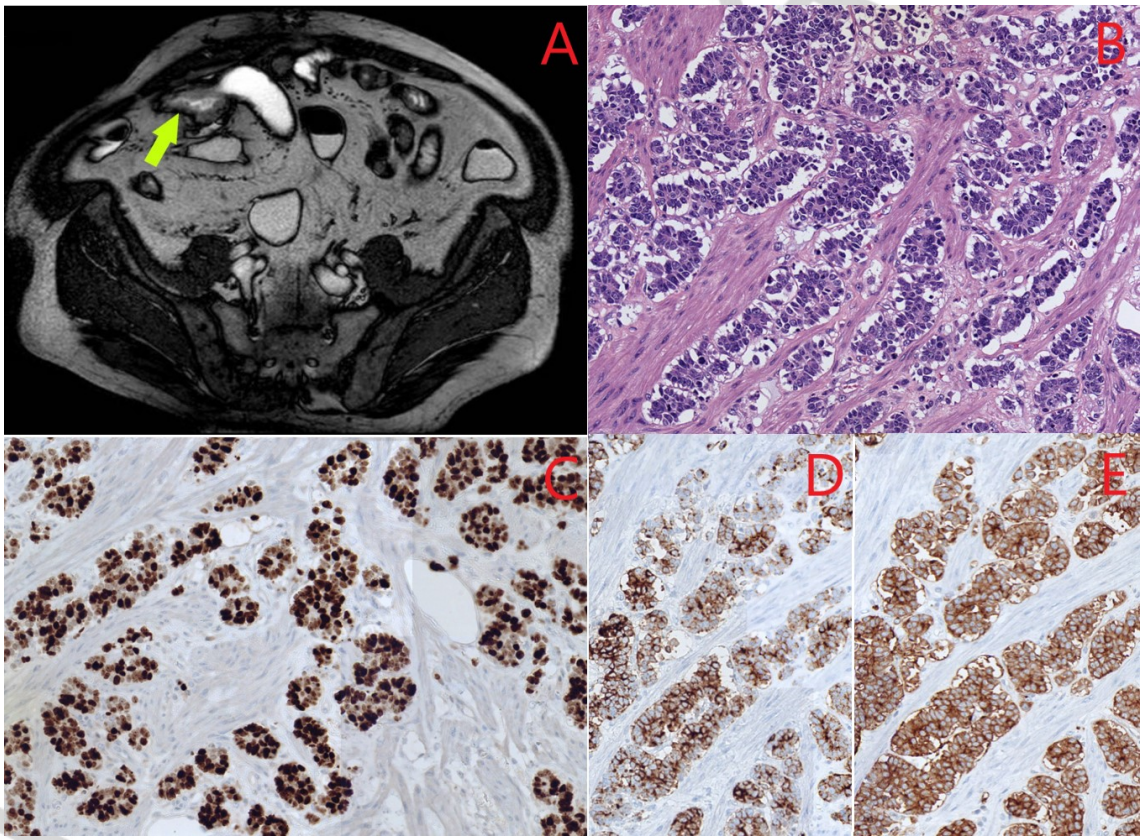

Figure 1. Abdominal MRI shows an asymmetric tumoral thickening of the ileum (A). The histopathological study of the resection specimen showed a well-differentiated tumor with trabecular and insular architecture (B). Ki-67 staining was detected in 80% (C) and immunochemistry staining was positive for neuroendocrine markers:

chromogranin (D) and synaptophysin (E).