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Lymphoepithelioma-like carcinoma of the duodenum: a very infrequent tumor

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Key words: Lymphoepithelioma-like carcinoma. Duodenum. Duodenal resection. Management.

Dear Editor,

Primary malignant tumors of the duodenum represent only 0.3% of malignant neoplasms of the gastrointestinal tract and adenocarcinoma is the most frequent histological type (1). Lymphoepithelial carcinoma is a histologically undifferentiated and rare neoplasm. It is characterized by a large reactive lymphoplasmacytic infiltrate in the stroma that usually presents with islets of undifferentiated cells, which are distinctively positive for Epstein-Barr virus (EBV). Lymphoepithelioma-like carcinomas (LELC) were first described in the nasopharynx (2) and mostly appear in the respiratory tract. Although isolated cases have been described at different levels of the gastrointestinal tract, this is the first case described in the duodenum.

Case report

A 77-year-old male presented with chronic microcytic anemia. A colonoscopy was normal and an esophagogastroduodenoscopy identified a neoformation in the third duodenal portion. The computed tomography (CT) reported a parietal thickening of the duodenum and surgical pathology reported a duodenal cavum LELC (T2N0M0).
Immunohistochemistry showed a conserved expression of MLH-1, MSH-2 and MSH-6 with EBER+ in 100% of the tumor cells (Fig. 1).

Discussion
According to the literature of LELC in the gastrointestinal tract, the association with EBV is present in the stomach (3) and ileum (4), but not in colon (5) or esophagus (3). In addition to EBV, LELC has also been associated with ulcerative colitis and Crohn’s disease (both ruled out in this patient). The treatment is mainly surgical. Adjuvant chemotherapy and radiotherapy is usually not necessary due to an early stage diagnosis, although it is indicated when the tumor develops in the stomach (3). Despite a lack of adjuvant therapy, the prognosis is better than in other histological types of tumors at the same location. The explanation seems to lead to the characteristic lymphoplasmacytic infiltrate of LELC, which may have a key role in the oncological outcome. More studies are needed to establish the role of chemotherapy, radiotherapy and the lymphoplasmacytic infiltrate in the prognosis of the disease.

Supported by Hospital Clínico Universitario Virgen de la Arrixaca, Murcia, Spain.

Informed consent statement: consent was obtained from the patient for publication of this report and any accompanying images.

References

Fig. 1. A and B. Infiltration of the muscular layer itself (A: HE, panoramic) reaching the fat (B: HE 40x). C. The tumor is composed of multiple nests separated from each other by an abundant lymphoid infiltrate (HE 100x). D. The tumor cells were intensely positive for EBER-1 and no loss of expression was observed for the microsatellites instability markers, PMSE and MSH6 (100x).