Ileal carcinoid tumor within Crohn’s disease

Key words: Carcinoid. Small-bowel neoplasma. Crohn.

Dear Editor,

We present a rare case combining Crohn’s disease with intestinal carcinoid recently treated at our Division of Colorectal Surgery.

Case report

A 51-year-old female with a long history of Crohn disease (CD) and ileocolic resection in the past, receiving medical treatment with mesalazine and azathioprine, was admitted reporting daily colicky abdominal pain, diarrhea, and asthenia. Evaluation with abdomen multislice computed tomography (MSCT) demonstrated huge adenopathies located at the neoileal mesentry (Fig. 1A), which were ecoguide biopsied. Colonoscopy ruled out colorectal carcinoma and test revealed high values of CEA in blood, 5HIIA in urine, with a completely normal value of the beta-2 microglobulin. Subsequent OctreoScan confirmed abnormal mesenteric adenopathies tracer accumulation, with a discrete focus in distal ileum. Biopsy was consistent with lymph node metastasis of typical carcinoid tumor.

The patient underwent scheduled laparotomy with adenopathic conglomerate resection, located in neoileal meso (Fig. 1B). The primary tumor, a 0.5 cm lesion located in ileum, 15 cm proximal to ileocolic anastomosis, was also resected. Pathology confirmed moderately differentiated neuroendocrine tumor (KI < 1 %).

Fig. 1. A. Intravenous contrast portal-phase MSCT. Solid masses with nodular morphology located in mesenteric root, with heterogeneous contrast enhancement and hypodense central area that suggests necrosis. B. Surgical view: Locoregional adenopathies located at the neoileal mesentery.
Discussion

Malignant tumors of the small bowel are extremely rare, being the major histological subtypes: Adenocarcinoma, lymphoma and carcinoid tumor. Patients with long standing inflammatory bowel disease have an increased risk, which is related to two main factors: the longer duration of the inflammation and the site and severity of the disease.

The association of CD and adenocarcinoma of the small intestine has been well established by several studies. On the other hand, malignant carcinoid of the small intestine associated with CD is rare. Higashi et al reported that out of the 286 cases of CD examined, there were 13 concomitant cases of malignant disorders. Among them, six cases had colorectal cancer, while only one of the cases had rectal carcinoid (1).

Last studies about carcinoid tumors found in patients with inflammatory bowel disease have come up to the question of whether there is an association between the pathogenesis of these tumors arising from a background of active CD and the disease itself (2).

It is believed that chronic inflammation, caused by proinflammatory cytokines (TNFα, IFNγ, IL 2), lead enteroendocrine cells to hyperplasia and carcinoid transformation over a 10-year period (3). The finding of the tumor in an area of uninflamed intestine can be explained by the healing effect that immunomodulator therapy has over the mucosa or due to the effect of distant proinflammatory mediators, rather than a local inflammatory effect from adjacent CD (4).

Some authors do not believe that CD is a causative factor for carcinoid tumors since almost all cases were found incidentally after surgery for inflammatory bowel disease. Others argue that the coexistence may be under-reported (5). The diagnosis of carcinoid tumor poses a challenge, and it is rare before surgery because both conditions can mimic one another as far as clinical symptoms and diagnostic tests are concerned.

In general, the presence of ileal CD refractory to medical therapy should alert us to the possibility of a small-bowel neoplasm.

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References