CASE NOTE

Incidental malignant carcinoid within Crohn ileitis

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The association of Crohn disease and adenocarcinoma of the small intestine and colon has been well established. However, malignant carcinoid of the small intestine associated with Crohn disease is rare. We describe the case of a patient with an incidental malignant carcinoid found within Crohn ileitis.

CASE REPORT

A 43-year-old male smoker presented with a long history of ileal Crohn disease and postprandial abdominal pain. Colonoscopy showed patchy colitis consistent with Crohn disease. Steroid therapy had failed to improve his symptoms, and his symptoms had progressed during the year he was on azathioprine. Small bowel follow-through and a computed tomography (CT) scan showed a partial obstruction in the distal ileum, possibly at several points. We also found gallstones, but a hepatobiliary iminodiacetic acid scan showed normal gallbladder emptying.

The patient’s internist referred him to general surgery for possible laparoscopic cholecystectomy, as his symptoms were predominantly postprandial. The patient was initially unwilling to undergo an ileal resection but agreed to a cholecystectomy.

We performed the cholecystectomy without complications, but it did not improve his symptoms. The pathology investigation showed cholelithiasis and chronic cholecystitis. Finally, after the patient’s symptoms continued to progress and he began losing weight, he agreed to a bowel resection. Interestingly, his pain did respond partially to pregabalin (150 mg at night).

We performed another colonoscopy, as he had not had one in over a year, and this showed no active colitis. We performed an ileocolectomy and primary anastomosis, including the cecum and 40 cm of ileum. There were 3 distinct strictures grossly consistent with Crohn disease. The patient recovered without incident.

Pathology results showed classic findings of Crohn disease, including patchy ulcerations, transmural inflammation, hypertrophy of the muscularis mucosae and muscularis propria and submucosal abscesses. However, despite there being no gross evidence of a tumour, some sections showed irregular glands and nests of neuroendocrine-appearing cells in acinar and trabecular arrangements invading the muscularis propria and extending through the serosa into the peri-ileal fat (Fig. 1). Seven lymph nodes were negative for cancer. Initially, we thought our patient had an adenocarcinoma of the small bowel, but immunohistochemical stains were positive for cytoplasmic chromogranin and synaptophysin, as well as pancytokeratins (AE1/AE3), which we would expect to be positive in carcinoma and carcinoid tumours (Fig. 2). We diagnosed invasive carcinoid tumour arising from a background of Crohn ileitis.
DISCUSSION

In 1986, there were only 9 reported cases in the world literature of carcinoid tumours coexisting with Crohn disease. By 1997, this number had increased to only 16. In 10 of 16 cases, carcinoid tumours were found incidentally after bowel resection, remote from the area of active Crohn disease. There have been only 3 previously reported cases of malignant carcinoid coexisting in the same segment as Crohn ileitis.

Some authors do not believe that Crohn disease is a causative factor for carcinoid tumours, whereas others argue that the coexistence may be under-reported, since these 2 lesions can mimic one another.

Our patient’s case involved an invasive carcinoid with no gross evidence of a tumour arising from a background of active Crohn disease. We believe this is evidence that Crohn disease may be a predisposing risk factor for malignant small bowel carcinoid, just as it is for the much more prevalent adenocarcinoma.

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References