A rare intestinal malignancy: mantle cell lymphoma

Approximately 30% of primary extranodal lymphomas occur in the gastrointestinal tract. Most of these present as single lesions, although gastrointestinal polyposis occurs in some 10% of cases, including conditions such as multiple lymphomatous polyposis (MLP) and immunoproliferative small-intestinal disease (IPSID) [1]. MLP is a rare intestinal malignancy characterized by the presence of numerous gastrointestinal polypoid lesions of malignant lymphoma. It is a non-Hodgkin’s B-cell lymphoma that is the gastrointestinal counterpart of mantle-cell lymphoma [2]. The polyps range from a few millimeters to several centimeters in size and involve the small and large bowel in 80–90% of cases and the stomach or duodenum in 50% of cases [3]. The condition often shows extra-abdominal dissemination, especially to peripheral lymph nodes.

A 68-year-old man presented with a history of abdominal pain, occult lower gastrointestinal bleeding, loss of weight, sideropenic anemia, and fever. Colonoscopy revealed a large number of different-sized polyps (Figure 1). Abdominal ultrasound showed enlarged retroperitoneal lymph nodes. Chest and abdominal computed tomography (CT) also demonstrated lymphoid dissemination. Gastroscopy and CT enteroclysis did not show polypoid lesions. Histopathological examination of the large-bowel polyps confirmed lymphocytes with irregular nuclei. Immunohistochemistry (DAKO/LSAB+DAB) of tissue samples showed CD20, CD5, CD43, and cyclinD1 positivity, and CD79-alpha, CD3, and cytokeratin negativity (Figure 2). Mantle-cell lymphoma was diagnosed on the basis of these data.

The patient underwent four cycles of chemotherapy with a protocol using cyclophosphamide, hydroxydaunomycin, Oncovin (vincristine), and prednisone (CHOP) plus rituximab (Mabthera). The control colonoscopy showed that the polyps had disappeared. At the time of writing, the patient had been asymptomatic and without any abnormal laboratory values for a 12-month follow-up period.

This case represents a rare form of colonic polyposis. Combined chemotherapy was successful and the change in the endoscopic appearance was spectacular. In the management of such rare cases, we recommend careful follow-up of patients by gastroenterologists and oncologists. In case of relapse, repeated chemotherapy or bone-marrow transplantation is needed.

References

1 Foss HD, Stein H. Pathology of intestinal lymphomas. Recent Results Cancer Res 2000; 156: 33–41

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