A 48-year-old man was admitted to our hospital because of repeated episodes of epigastralgia. Endoscopy showed multiple whitish granules extending from the 2nd to 3rd portion of the duodenum. Biopsy specimens showed well circumscribed follicles with a monotonous population of predominantly small cleaved cells that were positive for CD20, CD10 and BCL-2, but negative for CD5. A full staging study showed no abnormalities. The tumor was finally diagnosed according to the WHO classification as a stage I follicular lymphoma (FL), grade 1, of the duodenum and subsequently received irradiation to the involved area. After 3 years of follow-up, he is still in complete remission. Because FL arising in the duodenum has recently reported with increasing frequency, patients with multiple granules in the duodenum should be examined carefully.

Key words: follicular lymphoma, duodenum, radiation therapy, gastrointestinal tract

(Introduction)

The gastrointestinal (GI) tract is the most common extranodal site of origin for non-Hodgkin’s lymphomas (NHLs), accounting for approximately 40% of all extranodal primary NHLs. The stomach is the most common primary site for NHL, followed by the colorectal region and the terminal ileum (1). However, the duodenum is rarely involved (2, 3).

Follicular lymphoma (FL) is a neoplasm of follicle center B-cells and is one of the most common subtypes of NHL. Most patients with FL present with nodal involvement, and extranodal presentation is uncommon. Although FL of the GI tract represents only about 1-3.6% of all GI tract lymphomas (3-5), FL arising in the duodenum has been reported by occurring with increasing frequency (3, 6), and in fact recently there have been numerous case reports on this disease (7-11). Here, we present a case of stage I primary FL arising in the duodenum, which was diagnosed by biopsy specimens and treated successfully with irradiation to the involved area.

(Case Report)

A 48-year-old Japanese man presented with a 4-week history of epigastralgia. Initial endoscopic evaluation at another institution showed only gastric erosion; there was no evidence of duodenal abnormality. He was treated with antacids and obtained temporary relief from the symptoms. Later, however, the epigastralgia recurred, and he visited our hospital. Interview revealed that the patient had had poorly controlled diabetes for the previous ten years. Physical examination revealed no abnormalities, and there was no evidence of hepatosplenomegaly or peripheral lymphadenopathy. All laboratory findings, including the levels of lactate dehydrogenase and soluble IL-2 receptor, were within normal limits. Upper GI endoscopy revealed multiple whitish granules extending from the 2nd to 3rd portion of the duodenum...
Figure 1. Endoscopic view of the duodenal follicular lymphoma. Multiple whitish granules extend from the 2nd to 3rd portion of the duodenum.

Figure 2. Histopathological examination of endoscopic biopsy specimens, showing a low-power view (A) and a high-power view (B). The biopsy specimens show well circumscribed follicles (A) composed of a monotonous population of predominantly small-cleft cells (B).

Figure 4. Nine months after the start of radiotherapy, endoscopic examination demonstrates disappearance of the granular lesions.

Biopsy specimens showed well circumscribed follicles composed of a monotonous population of predominantly small-cleft cells (Fig. 2) that were positive for CD20, CD10 and BCL-2, but negative for CD5. The margin of the lesion revealed by endoscopy corresponded completely to that observed by microscopy. Rearrangement of the immunoglobulin heavy chain gene was confirmed by Southern blot analysis using tumor samples. Therefore, the tumor was diagnosed as a grade 1 FL, according to the WHO classification (12). Hypotonic duodenography showed thick and meandering folds, measuring about 50 mm, extending from the 2nd to 3rd portion of the duodenum (Fig. 3). Other examinations, including computed tomography, gallium scintigraphy, colonoscopy, endoscopic retrograde cholangiopancreatography, and bone marrow aspiration, showed no abnormalities. On the basis of these findings, the FL was evaluated as stage I.

The patient subsequently underwent irradiation to the involved area consisting of 30.6 Gy delivered in 1.8-Gy fractions. Nine months later, endoscopy revealed disappearance of the granular lesions (Fig. 4) and lack of lymphoma cells in biopsy specimens. Multiple biopsies of the duodenal mucosa during repeat endoscopies failed to demonstrate the presence of lymphoma cells. There was no evidence of recurrence during the 3 years of follow-up.

Discussion

FL is a rare histological subtype among primary GI tract NHLs. Recently, however, there has been increasing evidence that the incidence of FL is unexpectedly high in the duodenum in comparison with other GI sites. In the series reported by Yoshino et al (3), 5 of 8 cases of GI tract FL arose in the duodenum, and in another series reported by Shia et al (6), duodenal FL accounted for 10 of 26 cases of FL of the GI tract.

The clinical course of FL and treatment strategies differ according to the extent of disease (i.e., stage) at presentation. Although initial complete remission may be achieved by intensive therapies such as high-dose chemotherapy and/
or rituximab administration or chemo/radiotherapy followed by stem cell transplantation, patients with disseminated (stage III and IV) FL usually relapse and have difficulty in achieving complete cure, whereas those with localized (stage I and II) FL are usually treated by radiation therapy or surgical resection and/or chemotherapy and often show long survival (13). Although the majority of cases of primary duodenal FL are treated by surgical resection, some are diagnosed only after surgical resection because of the difficulty in diagnosing FL from biopsy specimens (3). Therefore, for management of this disease, it is essential to make an accurate diagnosis and carry out accurate staging. The present case was diagnosed as grade 1 FL from biopsy specimens, and the margin of the lesion revealed by endoscopy corresponded completely to that observed by microscopy, i.e. it was judged to be a stage I primary FL extending from the 2nd to 3rd portion of the duodenum.

In this case, radiation therapy was chosen because surgical resection was considered to carry a high risk due to the patient’s poorly controlled diabetes, and chemotherapy was not considered appropriate because the FL was localized. He received irradiation to the involved area consisting of 30.6 Gy delivered in 1.8-Gy fractions, without any adverse events, and has since been in complete remission for the last 3 years. Radiation therapy seems to be a safe and effective treatment modality for primary duodenal FL. However, Mac Manus et al reported that a few patients with localized FL suffered relapse, even decades after radiation therapy (14). Therefore, patients with FL need to be followed for as long as possible because of the risk of relapse.

In conclusion, we have reported a case of stage I primary FL arising in the duodenum, which was diagnosed by endoscopic biopsy and treated successfully with irradiation to the involved area. Patients with multiple granules in the duodenum should be examined carefully, especially in view of the fact that FL arising in the duodenum has recently been reported with increasing frequency.

References


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