Esophageal Involvement in Lymphoma

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Esophageal involvement by lymphoma in three patients, two with non-Hodgkin’s lymphoma and one with Hodgkin’s lymphoma are reported. In all three patients, there was discrete involvement of the esophagus not directly contiguous with the stomach. Esophageal involvement by lymphoma either as a primary disease or as manifestation of a disseminated disease is distinctly uncommon.

INTRODUCTION

The esophagus is an unusual site for lymphoma, where it is reported to occur in less than 1% of the cases even when the initial clinical manifestations of lymphoma occur in the gastrointestinal tract (1). We report three unusual cases of lymphoma with esophageal involvement. These cases are unique in the sense that despite extensive esophageal involvement the two cases with non-Hodgkin’s lymphoma had no esophageal symptoms. The third patient with Hodgkin’s disease developed severe esophageal symptoms 9 years after initial diagnosis and was proved to have Hodgkin’s disease of the esophagus while the remainder of the gastrointestinal tract was uninvolved.

CASE REPORTS

Case 1

A 41-year-old man was well until 1981 when he developed gradual weakness and lethargy which were attributed to mild anemia. No further work up was pursued until October 1982, when an enlarged spleen and leukopenia (white blood cells 2200/mm³) were found. A splenectomy was performed but no diagnosis was made. In March 1983 he developed cervical and axillary lymphadenopathy. A lymph node biopsy revealed mixed small and large cell lymphoma (lymphocytic-histiocytic) which was immunophenotyped on a subsequent lymph node biopsy as a T-cell lymphoma of suppressor cell type (Lennert’s lymphoma) (6, 7). CT scan showed paraaortic and pelvic lymphadenopathy. A bone marrow biopsy revealed lymphomatous involvement. Cerebrospinal fluid examination by lumbar puncture was normal. The patient was treated with chemotherapy consisting of Vincristine, Prednisone, and high dose Adriamycin (doxorubicin hydrochloride) in three courses. An abdominal CT scan following completion of initial chemotherapy revealed significant resolution of abdominal and pelvic lymphadenopathy. During the next 4 months due to fluctuant clinical course he received several courses of chemotherapy but continued to have fever. He was admitted for work up of refractory fever in January 1984. Abdominal CT scan, Gallium scan, and all cultures were negative for organisms. An upper gastrointestinal series revealed three discrete large ulcers in the esophagus on a background of normal mucosa (Fig. 1A). The stomach showed thickened mucosal folds. Endoscopic biopsy of the esophageal ulcers revealed lymphomatous infiltration similar to that found in the lymphnode biopsies (Fig. 1B). Biopsy of the stomach revealed no evidence of lymphoma. Biopsy of oral ulcers revealed lymphomatous infiltration. During the next few weeks the patient developed neck pain, as well as swelling of his mouth, face, and neck. His respiratory status and general condition continued to deteriorate and he died in February 1984.

Case 2

A 74-year-old white woman admitted for abdominal complaints and 20-lb weight loss was found to have a circumscribed ulcerating mass in the antrum of the stomach in January 1981. Biopsies of this lesion revealed diffuse histiocytic lymphoma. She underwent partial gastrectomy with Billroth I gastroduodenostomy and received 4000 rads to the tumor bed postoperatively. The patient did extremely well for 2½ years with minimal discomfort until approximately 4 months before admission in August 1983, she developed early satiety, dysphagia, nausea, vomiting, recurrent pyrosis, and a 10-lb weight loss.

An upper gastrointestinal examination revealed a polyloid eccentric mass in the distal esophagus immediately above the gastroesophageal junction (Fig. 2A), as well as a gastric ulcer with thickened folds in the gastric remnant. A CT scan revealed thickened wall of the distal esophagus (Fig. 2B). No paraaortic or pelvic lymphadenopathy was seen. Endoscopic biopsy revealed large cell histiocytic lymphoma involving the distal esophagus and the stomach. She was treated with chemotherapy. When last seen in January 1985 she had
no gastrointestinal symptoms and her disease was in remission.

Case 3
A 28-year-old man was diagnosed as having Hodgkin’s disease by cervical lymphode biopsy in 1960. He received radiation to the neck, axilla, and mediastinum at various times over the next 6 years. In 1969, 9 years after initial diagnosis of Hodgkin’s disease he began to experience dysphagia which progressed over the next several months and in May 1970 he was able to swallow only liquids with considerable substernal pain. He had 30-lb weight loss during the previous 2 months. An upper gastrointestinal series revealed smooth narrowing of the distal esophagus with moderate proximal dilatation (Fig. 3A). An endoscopic biopsy from this lesion did not reveal any tumor. However, an open biopsy of the distal esophagus confirmed the presence of Hodgkin’s disease (Fig. 3B). A feeding gastrostomy tube was placed. The patient received 2200 rads to the esophagus and chemotherapy. Two years later in 1972 he presented with malfunctioning gastrostomy tube and few constitutional symptoms. An upper gastrointestinal series showed complete obstruction of the esophagus and a probable mass at the gastroesophageal region distorting the fundus of the stomach. He received further chemotherapy and died 3 years later in January 1975.

DISCUSSION
Lymphomatous involvement of the gastrointestinal tract occurs in about 10–20% of all lymphomas (1–5, 8–11). The gastrointestinal tract may be involved primarily or as a secondary late manifestation of the disseminated disease (3). The most common site in the gastrointestinal tract for lymphoma is the stomach (48–50%), followed by the small bowel (30–37%), the ileoceleal region (12–13%), and the colon (10–12%) (2). Primary involvement of the esophagus by lymphoma is very rare (about 1%). Our literature search indicates several publications, mostly case reports of esophageal involvement of Hodgkin’s lymphoma (12–21) and non-Hodgkin’s lymphoma (12, 21–30). In one large series of 1269 cases of lymphoma, only 19 had disease in the esophagus and none had esophagus as a presenting site (1). When lymphoma is found in the esophagus, it usually affects the distal portion with contiguous involvement of the gastric fundus (23–31). Research of
lymphoma cases indexed at the surgical pathology files of the University of Michigan Hospitals for the years 1961–1983 revealed 123 cases with gastrointestinal lymphoma. No other cases with esophageal involvement by lymphoma were found except three reported herein.

The lymphomatous change starts in the submucosal lymphoid patches and may produce a wide spectrum of radiological features in the gastrointestinal tract. The involved segment may manifest varying size nodules, enlarged folds, polypoid mass, ulceration, stenosis, varicoid appearance due to diffuse submucosal infiltration, and aneurysmal dilatation. Except for aneurysmal dilatation all other features of lymphoma have been reported in the esophagus (23, 27). Spontaneous perforation of the esophagus (18) and malignant tracheoesophageal fistula have also been reported (32). Salem et al. (33) reported a case of histiocytic lymphoma with initial presentation of portal hypertension and bleeding esophageal varices due to compression of the portal vein by the lymphomatous enlarged nodes.

In our case 1 the presentation of three discrete ulcers in the esophagus on a background of normal mucosa is most unusual. It is believed that in case 2 there was synchronous development of lymphoma in the esophagus and stomach rather than transcardial extension of gastric lymphoma. These two patients with non-Hodgkin’s lymphoma had no esophageal symptoms despite extensive esophageal involvement. The third patient with Hodgkin’s lymphoma had significant odynophagia, due to desmoplastic response and a hard fibrotic stricture.

It has been suggested that if esophageal lymphoma is present without gastric involvement an extra-esophageal site of disease is usually identified (24, 25). Primary versus secondary esophageal involvement by lymphoma is difficult to determine, because in most instances other parts of the gastrointestinal tract are usually involved. The most frequent complications of esophageal involvement by lymphoma are hemorrhage, obstruction, and perforation with esophagomediastinal or esophagotracheobronchial fistula. Invasion of the esophagus is usually considered a poor prognostic indicator. Ehrlich et al. (19) reported in their series two patients with reticulum cell sarcoma and seven with Hodgkin’s disease who died within 3 and 7 months,
respectively, from the onset of esophageal involvement. Our case 1 died within 1 month of the detection of esophageal disease. It was obviously a terminal event of a widespread disease. Case 2 is alive 18 months after detection of esophageal disease and has responded well to combination therapy. The third case with Hodgkin's disease survived 5 years after detection of esophageal disease.

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REFERENCES

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