A 75-year-old man was admitted due to a 6-month history of 15 kg weight loss, diarrhea and fever. The physical examination showed cachexia. Important pathological laboratory values were: erythrocyte sedimentation rate (ESR) 87/96 mm, lactate dehydrogenase (LDH) 597 U/l (rising to 2030 U/l within 20 days), and albumin 2.2 g/dl. The patient had macrocytic anemia, and gliadin and endomysial antibody tests were positive.

Upper endoscopy revealed a bulging tumor in the middle part of the esophagus (Figure 1) and mucosal atrophy in the duodenum. Microscopic examination of the esophageal biopsies (Figure 2) showed that the lesion was an anaplastic large-cell lymphoma. The histological findings in the duodenal biopsies were consistent with (previously undiagnosed) celiac disease. Immunostaining of the duodenal intraepithelial lymphocytes showed the aberrant phenotype CD3⁺CD8⁻, which was the same as in the esophageal large-cell lymphoma. In addition, T-cell receptor γ-chain polymerase chain reaction (PCR) testing disclosed the same monoclonal rearrangement in both the duodenal and the esophageal biopsy. A bone-marrow biopsy was positive for anaplastic large-cell lymphoma. One week after diagnosis, the patient died of pneumonia and septic complications.

Intestinal T-cell lymphoma (ITL) usually arises as a complication of celiac disease, and has therefore been referred to as “enteropathy-type T-cell lymphoma” [1]. To the best of our knowledge, ITL presenting as an esophageal mass has not previously been reported. It may be hypothesized that in this case, the gluten-triggered intestinal inflammatory process became self-sustaining, converted to a monoclonal T-cell proliferation, and on dissemination developed into overt large-cell lymphoma, presenting clinically as a mid-esophageal mass. The clinical course of patients with ITL is very unfavorable, due to immediate complications arising from peritonitis and malnutrition and later from progressive disease. Roughly half of the patients are amenable to chemotherapy. The 5-year survival ranges from 8% to 25% [2–5].

References


Corresponding Author

T. Weber, M.D.
1. Interne Abteilung
Allgemeines Krankenhaus
der Barmherzigen Schwestern
Grieskirchnerstrasse 42
4600 Wels
Austria
Fax: +43-7242-415 3992
E-mail: webertom@aon.at