A 75-Year-Old Man with Fever, Diarrhea, Weight Loss and a Mid-Esophageal Mass

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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].

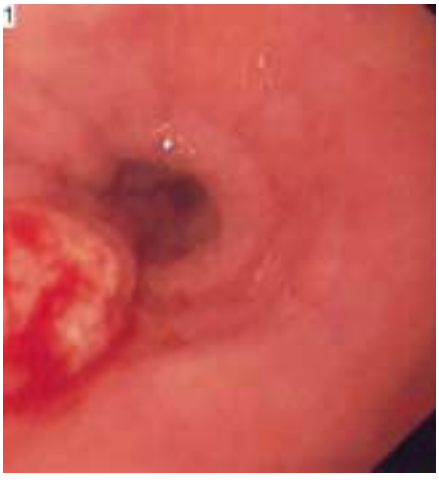

Figure 1 Endoscopic image of the middle part of the esophagus. There is a bulging tumor 10 × 3 mm in size, covered with a whitish layer

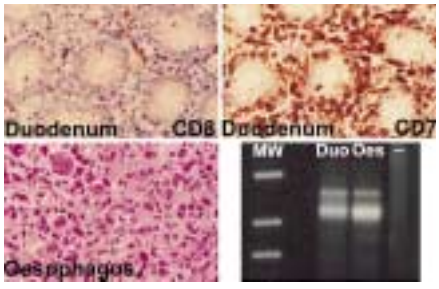

Figure 2 Histological, immunophenotypic, and genotypic analyses of duodenal intraepithelial lymphocytes (IEL) and esophageal large cell lymphoma. All duodenal IEL stain for T-cell associated antigen CD7, but only a few scattered lamina propria lymphocytes are reactive to CD8, indicating an aberrant CD8-phenotype (upper pair of images). The esophageal tumor is composed of anaplastic large cells. Both lesions show the same biallelic monoclonal T-cell receptor-γ-chain gene rearrangement suggesting that the esophageal lymphoma has emerged from the abnormal duodenal IEL population (lower pair of images)

References

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