

Enteropathy-associated T cell lymphoma as a complication of longstanding celiac disease

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ABSTRACT

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CASE REPORT

A 59-year-old female presented with weight loss, night sweats, and diarrhea since four months. Stool examination, serologic testing, esophagogastroduodenoscopy (EGD), colonoscopy (C-scope), and computed tomography (CT) scan of chest and abdomen were non-contributory. Diagnosed with celiac disease at 51, she had had no prior digestive symptoms despite poor adherence to a gluten free diet and persistently positive anti-tissue transglutaminase antibodies. Physical examination revealed cachexia, widespread erythematous maculopapular skin lesions, and no palpable lymphadenopathy. Laboratory examination showed hemoglobin 9.8 g/dL, mean cell volume 92 μm^3 , white blood cell count 5500/ μL (lymphopenia at 300/ μL), and serum albumin 2.2 g/dL.

The PET scan showed intense FDG uptake in a left tonsillar mass with multiple FDG-avid subcutaneous lesions, and bowel and mesenteric involvement (Figure 1). The left tonsil and one of the cutaneous lesions were biopsied, revealing a CD8+ T-lymphoid cell infiltrate, most consistent with T cell lymphoma showing a CD4 negative, CD8 positive, and CD 56 negative cytotoxic phenotype. Balloon enteroscopy revealed a mosaiform small bowel mucosa with multiple clean-based ulcers throughout the jejunum. Biopsies were consistent with the tonsillar findings. A few days following admission

the patient underwent a laparotomy for small bowel perforation, and was subsequently transferred to the intensive care unit where her course was complicated by recurrent gastrointestinal bleeding, septic shock and respiratory failure, leading to death.

DISCUSSION

Celiac disease (CD) is an immune-mediated enteropathy triggered by ingestion of gluten in genetically

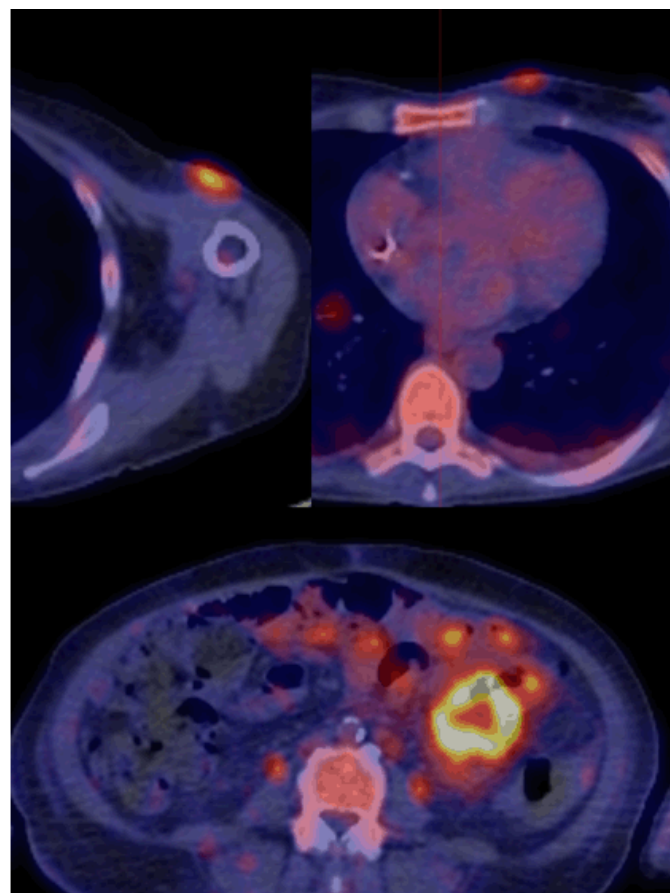


Figure 1: 18-FDG PET scan showing multiple FDG-avid subcutaneous lesions as well as bowel and mesenteric involvement.

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susceptible individuals. It is a common disorder affecting approximately 0.5–1% of Caucasians [1]. Overall, 2–10% of patients with CD develop (RCD) [2]. Refractory celiac disease can be divided into types I (RCD I) and II (RCD II). About half of RCD II patients develop enteropathy-associated T cell lymphoma (EATL) [3]. RCD II is therefore considered an EATL precursor lesion. Although rare, EATL is one of the main causes of death in patients with symptomatic CD diagnosed as adults. In patients with CD presenting with fever, night sweats, pruritus, significant unexplained weight loss, prolonged diarrhea, anorexia, overt or occult gastrointestinal bleeding, abdominal pain, and bowel obstruction, EATL should be sought [1, 3].

Multiple endoscopic or imaging techniques are available to visualize the entire small bowel such as capsule endoscopy, CT or magnetic resonance imaging enterography, deep enteroscopy, and 18F-FDG PET scan. The latter two techniques are of great utility to exclude malignancy in CD patients presenting with alarm features.

Abdominal CT scan has shown limited effectiveness in detecting EATL and distinguishing it from uncomplicated refractory celiac disease. Findings of bowel wall thickening, lymphadenopathy, intussusception and hyposplenism raise suspicion for RCD II and EATL. Apart from evidence of metastatic disease there are no well-described CT findings that will distinguish EATL from RCD II. Moreover, the role of radiologic imaging may be limited in diagnosing EATL as the neoplastic changes may be restricted to the epithelial layer of the small bowel, even when the lymphoma affects the whole small intestine [4].

The PET scanning displays higher sensitivity and specificity for the detection of EATL when compared with CT scan [5], but yields false positive results due to increased uptake in inflammatory tissues as is seen in non-refractory CD and RCD. However, the standard uptake value is lower than in EATL (0.0–4.6 vs 6.4–8.0 SUV), making PET a reliable method for identifying EATL [6]. Although PET scanning cannot be a substitute for histologic examination in diagnosing EATL, it may guide histological sampling.

Endoscopic examination of the small bowel in patients with celiac disease permits direct visualization and biopsy. Using EGD yields a limited examination of the small bowel, whereas only the distal ileum can be reached with colonoscopy [7]. Deep enteroscopy can provide full-length examination of the small bowel, and the ability to both perform biopsies and deliver therapy [7].

Video capsule endoscopy (VCE) allows full examination of the small bowel and assessment of the extent of involvement. Mucosal flattening and ulcerations can be found in EATL, but are also seen in ulcerative jejunitis. Video capsule endoscopy may also yield false negative results, as EATL can present as a sub-mucosal mass. Thus, reliable detection using VCE may be difficult. Most importantly, detected lesions cannot be sampled at VCE for histological examination [8].

Once a diagnosis of EATL is confirmed, most patients undergo surgical debulking followed by chemotherapy that usually includes the administration of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP). If feasible, surgery aids in diagnosis, while also decreasing the risk of potentially lethal perforation following chemotherapy and tumor necrosis. Stem cell transplantation (SCT) may be considered in patients presenting with an overall good clinical state, although no randomized trials have been performed. Unfortunately, as 64% of patients are diagnosed with stage IV disease, the prognosis remains very poor with one and five-year survival rates of 31–39% and 8–20% respectively [1].

CONCLUSION

We present a 59-year-old female with celiac disease of eight years duration who developed weight loss, night sweats, and diarrhea since four months. Biopsies at enteroscopy confirmed an enteropathy-associated T cell lymphoma, noted in 50% of patients with refractory celiac disease type II. As most, she had advanced stage IV disease; she died of small bowel perforation.

Keywords: Celiac disease, Enteropathy-associated T cell lymphoma, Enteroscopy, Positron emission tomography scanning

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Melika Hosseina – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising

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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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