Enteropathy-associated T-cell lymphoma: involvement of the gastrointestinal tract from the duodenum to the rectum

Enteropathy-associated T-cell lymphoma (EATL) is a rare type of non-Hodgkin’s lymphoma that is commonly associated with celiac disease. The disease is very aggressive with a poor prognosis, and no standardized treatment protocol has been established [1]. An early diagnosis and effective therapy may not be achieved because of the nonspecific clinical and endoscopic findings [2]. The radiologic features of the disease include wall thickening, ulceration, and perforation of the jejunum [3]. 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography/computed tomography is a useful tool for the staging, management, and prognostication of T-cell lymphoma [4]. Here, we present a unique case of EATL with diffuse involvement of the intestine and colon. A 41-year-old woman was admitted to the hospital with abdominal pain and vomiting in September 2014. Her medical history included celiac disease. Free intraperitoneal fluid and liver heterogeneity were detected by ultrasound. Computed tomography revealed multiple hypodense lesions without contrast enhancement in the liver and contrast-enhanced nodularity, which supported the diagnosis of peritoneal involvement.

Fig. 1  Histopathologic findings in a 41-year-old woman with enteropathy-associated T-cell lymphoma (EATL). a Celiac disease. Non-neoplastic mucosa distant from an intestinal T-cell lymphoma shows villous atrophy, crypt hyperplasia, and an increase in cytologically unremarkable intraepithelial lymphocytes without evidence of lymphoma (hematoxylin and eosin [H&E] stain, original magnification × 100). b EATL shows ulcerated neoplastic colonic mucosa with infiltrating atypical lymphocytes. The tumor cells are medium to large transformed lymphoid cells with round or angulated vesicular nuclei, prominent nucleoli, and moderate pale-staining cytoplasm between colonic glands (H&E stain, original magnification × 200). c EATL atypical lymphocytes are positive for CD3 (immunohistochemical stain, original magnification × 200).

Fig. 2  Positron emission tomography/computed tomography (PET/CT) images. Axial PET/CT fusion (a), axial PET (b), and sagittal (c) and coronal (d) PET/CT fusion images. e Maximum-intensity projection image shows diffuse involvement of the entire gastrointestinal tract from the duodenum to the rectum with a high rate of 18F-fluorodeoxyglucose (18F-FDG) uptake and enlargement of the intestinal wall. 18F-FDG was evident in and around the liver.
tonitis carcinomatosa. The results of endoscopic and rectoscopy were compatible with gluten enteropathy. Owing to her worsening clinical condition, the patient underwent laparoscopy, which revealed multiple nodular peritoneal lesions. A biopsy revealed EATL (Fig. 1).

$^{18}$F-FDG positron emission tomography/computed tomography performed for staging showed wall thickening in the gastrointestinal tract and intense FDG uptake, beginning in the duodenum and extending to the rectum (Fig. 2). In addition, nodularity and a high rate of $^{18}$F-FDG uptake were detected at peritoneal sites, and FDG uptake was increased in and around the liver. Repeat endoscopy and colonoscopy showed multiple nodular lesions in the duodenum and various locations within the colon (Fig. 3). The patient died 1 month after the initiation of chemotherapy.

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