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Case Report

Terminal Ileum Perforation Revealing an Enteropathy Associated T-Cell Lymphoma

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Abstract

Enteropathy Associated T-Cell Lymphoma (EATL) is a rare gastrointestinal non-Hodgkin's lymphoma. Complications are frequent, especially in type II EATL, such as intestinal obstruction, bleeding, or perforation. We describe in this case a small bowel perforation revealing an EATL.

Keywords: T-cell lymphoma; Enteropathy; Terminal ileum perforation

Introduction

Enteropathy Associated T-Cell Lymphoma (EATL) is a rare gastrointestinal non-Hodgkin's lymphoma [1,2]. This condition is revealed in a certain number of cases with perforation of the involved bowel, which worsens an already poor prognosis. We describe in this case a small bowel perforation revealing an EATL.

Case Report

A 64-year-old man presented to the emergency room after a sudden onset of abdominal pain. On examination, he had a fever (39°c), rebound tenderness in the right iliac fossa and tenderness in the hypogastrium. The laboratory tests showed leukocytosis (white blood cell count of 16,690/ml) and elevated C-reactive protein (119 mg/l). A CT-scan revealed free intra-peritoneal air with perforation of distal small bowel (Figure 1).

Emergent laparotomy was performed, which showed circumscribed peritonitis with pelvic abscess in the pouch of Douglas, secondary to a perforation of the distal ileum. This perforation was adjacent to an area of a macroscopic lymphomatous involvement. A 30 cm length of ileum containing the perforation was resected and a side to side anastomosis was performed.

Post-operative management was uneventful and the patient was discharged after recovery from the acute perforation. In the pathology report and on the basis of microscopic findings and immunochemistry, we made the diagnosis of EATL.

Discussion

Enteropathy Associated T-Cell Lymphoma (EATL) represents less than 1% of non-Hodgkin's lymphomas [1,2]. There are two types of EATL: Secondary EATL associated with Celiac Disease (CD). And de novo EATL in patients with no history of complicated CD. In secondary EATL, the diagnosis of CD is anteriorly established in 20 to 40% of cases [3,4]. Patients with gluten-sensitive CD can develop EATL in 7 to 10% of cases [1,5]. However, in case of refractory celiac disease, which does not respond to a Gluten-free regimen, the transition into EATL is more common and accounts for about 52% [5]. More recently, EATL was sub-classified into two types according pathological and phenotypic features [6]: type I EATL generally associated with CD and type II EATL generally not associated with CD.

The mean age at diagnosis is 64 years and approximately 65% of cases are diagnosed in men [7]. The main clinical symptom is abdominal pain which seldom motivates further investigations, especially when there are no other accompanying symptoms like fever or weight loss. Therefore, complications are frequent, especially in type II EATL, such as intestinal obstruction, bleeding, or perforation like our patient. In a large retrospective French study [8], including 37 patients, the disease was revealed in 32.4% of cases either with intestinal obstruction (9 patients), or perforation (3 patients). A Dutch retrospective study [5], including 93 patients, showed that emergent laparotomy after complications was conducted in 11 subjects. Peritonitis after bowel perforation presented 5 out of these 11 cases.

These complications develop on prior lesions as ulcers and strictures. In the previously mentioned French study [8], EATL was confined in the small bowel in 28 subjects. Multiple synchronous lesions were found in 54% of cases. Perforations involve generally the proximal jejunum, where jejunal ulcers due to EATL lesions predominate. Less frequently, this complication concerns the rest of the small bowel, like our case, or the colon [9].

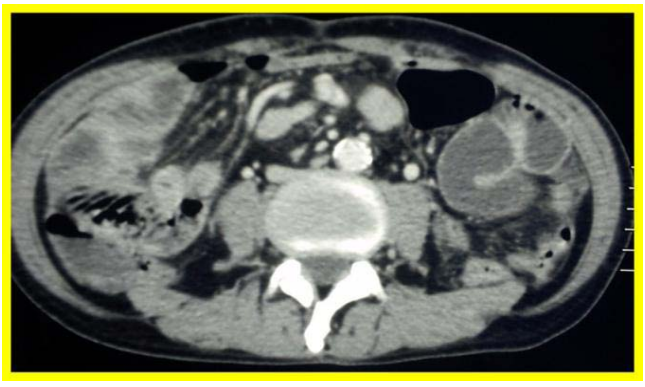

Figure 1: Thickened wall of terminal ileum.

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Diagnosis of EATL requires a biopsy, which can be obtained by endoscopy or surgery. Generally surgery is indicated because diagnosis could not be established with less invasive methods. Histology shows infiltration of the intestinal epithelium by lymphoid cells. These lymphocytes are variable in size and morphology in type I EATL and the immunophenotype shows CD3+, CD8−, and CD56−. Whereas in type II EATL, lymphocytes are monoclonal and the immunophenotype shows CD3+,CD8+, and CD56+ [10].

At present, there are no protocols for the diagnosis or treatment of EATL.

Treatment is mainly based on chemotherapy, like other lymphomas, with poor results. The role of elective surgery is however still not obvious.

In the French study, aggressive chemotherapy as well as serum albumin level >21.6 g/L were associated with better survival. In the same study, 25 patients underwent surgery and 22 had resection of the tumor. Results showed increased survival with debulking surgery in both univariate and multivariate analysis ($p<0.0001$ and $p<0.03$ respectively).

On the other hand, we noted in the literature a higher risk of perforation after the first cycle of chemotherapy [4], especially with type II EATL, which worsens the prognosis. This leads us to emphasize the importance of immunophenotype prior to chemotherapy, and raise the question whether we should consider an elective surgery before chemotherapy, to resect the tumor when it is localized.

In the era of evidence based medicine, dogmas should be erased, and more studies need to be conducted on the role of surgery in the management of EATL.

The prognosis of EATL is poor. One-year survival rate of the intestinal T-cell non-Hodgkin's disease is 33%, and five-year survival is 9% [3]. But, we need to lay stress on the difference in the prognosis between types of EATL. Better 5 year survival rate is noted with EATL associated with a CD responsive to a gluten free regimen. Also, needless to say that the earlier the diagnosis and treatment, the better the prognosis.

Conclusion

EATL is an uncommon lymphoma with frequent complications such as perforation or obstruction. They reveal the disease in about 50% of cases. Treatment involves chemotherapy and if it is associated with celiac disease, gluten should be suppressed from the diet. More studies need to be done in order to set the role of elective surgery in the management of this disease. Prognosis is poor, especially when the diagnosis is late. Therefore, we emphasize the need of a biopsy to diagnose EATL early and plan adequate approach in order to enhance outcomes.

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