ENTEROPATHY ASSOCIATED T-CELL LYMPHOMA – A RARE CAUSE OF ACUTE ABDOMEN

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CASE REPORT

Doi: 10.33695/rojes.v1i1.39
Accepted: 04.04.2021

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Abstract

Enteropathy associated T-cell lymphoma (EATL) is an extremely aggressive form of non- Hodgkin lymphoma. EATL is a rare form of peripheral lymphoma with a very poor prognosis and the five-year survival rate reported is between 11 and 20%. We present the case of a 33- years old male admitted for right iliac fossa pain and nausea. Blood test showed leukocytosis with neutrophilia. The conventional X-ray examination of the abdominal region showed a few "air-fluid" levels in the right flank and mesogastric area and the ultrasound found no particular alterations of the abdominal organs. These lead us to suspect an acute appendicitis and the patient was scheduled for emergency surgery. Intraoperative, we ascertain a perforated, stenotic jejunal tumor located at approximately 1 meter from the Treitz angle. A segmental enterectomy was performed with entero-enteral anastomosis end to end. The histopathological and immunohistochemistry examinations established the diagnosis of entheropathy associated T-cell lymphoma. The patient was discharged after a few days and started the proper adjuvant therapy. In most of the cases, the precise diagnosis is not facile, the patient usually presenting with intestinal haemorrhage, intestinal occlusion or perforation, the clinical presentation often misleading the medical team. EATL is usually a diagnosis of exclusion, other differential diagnoses being acute appendicitis, mechanical obstruction, intestinal bacterial infection and others.

Keywords: EATL, non-Hodgkin lymphoma, jejunal tumor, acute abdomen

Introduction

Enteropathy associated T-cell lymphoma is a rare and extremely aggressive form of non-Hodgkin lymphoma. EATL has a very poor prognosis, the five-year survival rate reported to be between 11 and 20 % [1][2]. Pre-operative diagnosis is difficult, most of the times EATL being a diagnosis of exclusion due to its clinical presentation which can vary from intestinal haemorrhage to intestinal occlusion or perforation [3]. The differential diagnosis from a clinical point of view includes acute appendicitis, mechanical obstruction, intestinal bacterial infections and others. Even though this type of small intestine

lymphoma in associated in 90% of the cases with celiac disease, there are patients with no signs or symptoms of malabsorption or gluten-sensitive enteropathy, the histopathological examination being the only investigation that can show signs of celiac disease clinical unapparent [4]. Most of the times, due to the acute complications that occur, the exploratory laparotomy and resection of the tumor is required, followed by immunohistochemistry and proper adjuvant therapy [5].

Case presentation

We present the case of a 33-years old male who came to the Emergency Department with right iliac fossa pain and nausea, without any other significant personal medical history. The symptoms appeared 7 days before and worsened in the past 24 hours prior to his admission

Clinical exam reveals diffuse abdominal pain both spontaneously and when palpated more accentuated in the right iliac fossa with positive Bloomberg sign in this region.

Initial blood test revealed leukocytosis (19310 /mm3) with neutrophilia (83,9 %). There were no other significant changes in the blood tests.

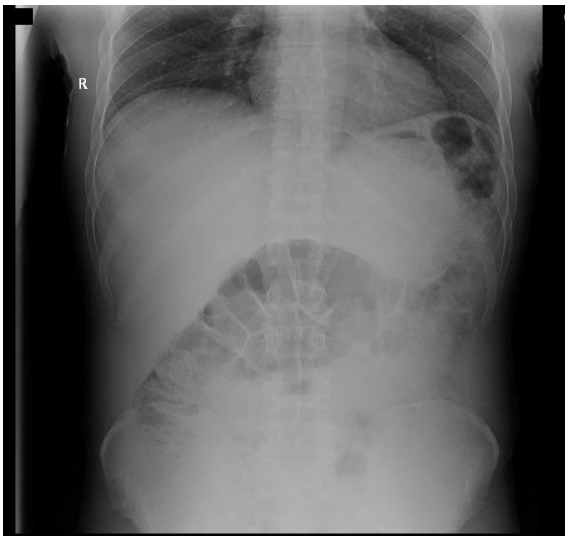

Figure no. 1 - "Air-fluid" levels in the right flank and mesogastric area

The first imaging technique used was abdominal ultrasound that found no particular alteration of the abdominal organs. The conventional X-ray examination of the abdominal region showed a few "air-fluid" levels in the right flank and mesogastric area (Figure 1). These lead us to suspect an acute appendicitis and the patient was scheduled for emergency surgery.

Intraoperative, we ascertain a perforated, stenotic jejunal tumor located at approximately 1 meter from the Treitz angle with an abscess located near the tumor (Figure 2 & 3). A segmental enterectomy was performed with entero-enteral anastomosis end to end.

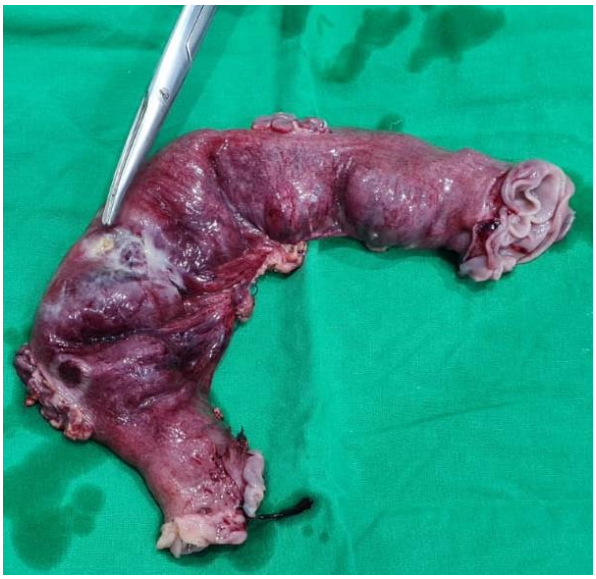

Figure no. 2 - The resected jejunal segment showing the perforated, stenotic tumor

The resected segment was sent for histopathological examination. The result describes extensive ulcerated area covered with fibrino-necrotic exudate and leukocytes; tumor proliferation with atypical pleomorphic large cells with large nuclei, prominent nucleoli and frequent atypical mitosis; the mucosa adjacent to the tumor proliferation has complete villous atrophy and crypt hyperplasia.

The immunohistochemistry examination shows positive results for EMA (Epithelial

Membrane Antigen), SATB2 (Special AT-rich sequence-Binding protein 2), CLA (Cutaneous Lymphocyte Antigen), CD3, MUM-1 (Multiple Myeloma Oncogene 1). The histopathological and immunohistochemistry examinations correlated with the clinical and paraclinical investigations conclude that the diagnosis is type I Enteropathy associated T-cell Lymphoma (EATL).

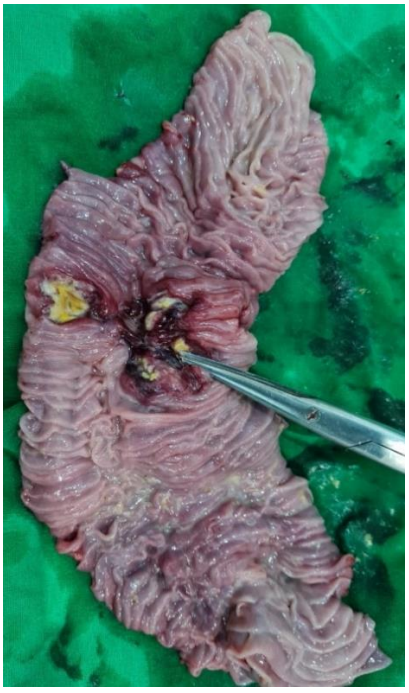

Figure no. 3 - The dissected jejunal segment showing the macroscopic ulcerated aspect of the tumor

The patient's evolution was favorable, the clinical symptoms disappeared and the blood tests normalized after a few days. The patient remained stable in the postoperative period and started adjuvant treatment with high-dose chemotherapy as recommended by the oncologist.

Discussion

Intestinal T-cell lymphoma is divided in two major groups: NK/T-cell lymphoma and enteropathy associated T-cell lymphoma according to World Health Organization classification [6]. EATL has two forms: type I is frequently associated with celiac sprue and express usually CD-30, type II is less frequently associated with celiac disease and is characterized by the expression of CD-56 [7]. EATL has an incidence of 0,1 per 100,000 per year, as some studies had shown [8] and because of its rarity there are no clear protocols regarding a specific treatment, even though this kind of lymphoma had been reported to an increase of incidence [9]. The CD-30 positive marker in the intraepithelial lymphocytes (IELs) has the worse prognosis, the expression of this marker being associated with the occurrence of overt lymphoma[10].

The symptoms that a patient with type I EATL may experience are extremely varied and nonspecific. They usually present with weight loss, abdominal pain, bowel movement changes or even acute abdomen[11]. The diagnosis in early stages is challenging due to these forms of presentation. The histopathological and immunohistochemistry examinations remains the gold standard in diagnosing enteropathy associated T-cell lymphoma [12-13].

Conclusions

There are many controversial opinions about the correct management of EATL patient. Even though the result of surgery are poor, the clinical presentation may impose an emergency intervention. In this kind of situation, the correct approach is additional systemic chemotherapy.

References

[1] P. Domizio, R. A. Owen, N. A. Shepherd, I. C. Talbot, and A. J. Norton, "Primary lymphoma of the small intestine. A clinicopathological study of 119 cases.," Am. J. Surg. Pathol., vol. 17, no. 5, pp. 429–442, May 1993, doi: 10.1097/00000478-199305000-00001.

[2] J. M. W. van de Water, S. A. G. M. Cillessen, O. J. Visser, W. H. M. Verbeek, C. J. L.

M. Meijer, and C. J. J. Mulder, "Enteropathy associated T-cell lymphoma and its precursor lesions," Best Pract. Res. Clin. Gastroenterol., vol. 24, no. 1, pp. 43–56, 2010, doi: https://doi.org/10.1016/j.bpg.2009.11.002.

[3] Z. H. Sun, H. M. Zhou, G. X. Song, Z. X. Zhou, and L. Bai, "Intestinal T-cell lymphomas: A retrospective analysis of 68 cases in China," World J. Gastroenterol., vol. 20, no. 1, pp. 296–302, 2014, doi: 10.3748/wjg.v20.i1.296.

[4] D. P. Arps and L. B. Smith, "Classic versus type II enteropathy-associated T-cell lymphoma: diagnostic considerations.," Arch. Pathol. Lab. Med., vol. 137, no. 9, pp. 1227–1231, 2013, doi: 10.5858/arpa.2013-0242-CR.

[5] A. Di Sabatino, F. Biagi, P. G. Gobbi, and G. R. Corazza, "How I treat How I treat enteropathy-associated T-cell lymphoma," vol. 119, no. 11, pp. 2458–2468, 2012, doi: 10.1182/blood-2011-10-385559.2458.

[6] J. W. Campo, E., Harris, N. L., Jaffe, E. S., Pileri, S. A., Stein, H., Thiele, J., & Vardiman, "WHO classification of tumours of haematopoietic and lymphoid tissues," 2008, [Online]. Available: http://apps.who.int/bookorders/anglais/detart1.jsp?codlan=1&codcol=70&codcch=4002.

[7] J. Delabie et al., "Enteropathy-associated T-cell lymphoma: Clinical and histological findings from the international peripheral T-Cell lymphoma project," Blood, vol. 118, no. 1, pp. 148–155, 2011, doi: 10.1182/blood-2011-02-335216.

[8] W. H. M. Verbeek, J. M. W. Van De Water, A. Al-Toma, J. J. Oudejans, C. J. J. Mulder, and V. M. H. Coupé, "Incidence of enteropathy--associated T-cell lymphoma: a nation-wide study of a population-based registry in The Netherlands.," Scand. J. Gastroenterol., vol. 43, no. 11, pp. 1322–1328, 2008, doi: 10.1080/00365520802240222.

[9] R. Z. Sharaiha, B. Lebwohl, L. Reimers, G. Bhagat, P. H. Green, and A. I. Neugut, "Increasing incidence of enteropathy-associated T-cell lymphoma in the United States, 1973-2008," Cancer, vol. 118, no. 15, pp. 3786–3792, 2012, doi: 10.1002/cncr.26700.

[10] I. N. Farstad et al., "Heterogeneity of intraepithelial lymphocytes in refractory sprue: Potential implications of CD30 expression," Gut, vol. 51, no. 3, pp. 372–378, 2002, doi: 10.1136/gut.51.3.372.

[11] J. C. Zhang, Y. Wang, X. F. Wang, and F. X. Zhang, "Type I enteropathy-associated T-cell lymphoma in the colon of a 29-year-old patient and a brief literature review," Onco. Targets. Ther., vol. 9, pp. 863–868, 2016, doi: 10.2147/OTT.S96745.

[12] J. Gale, P. D. Simmonds, G. M. Mead, J. W. Sweetenham, and D. H. Wright, "Enteropathy-type intestinal T-cell lymphoma: clinical features and treatment of 31 patients in a single center.," J. Clin. Oncol. Off. J. Am. Soc. Clin. Oncol., vol. 18, no. 4, pp. 795–803, Feb. 2000, doi: 10.1200/JCO.2000.18.4.795.

[13] M. Sieniawski et al., "Evaluation of enteropathy-associated T-cell lymphoma comparing standard therapies with a novel regimen including autologous stem cell transplantation," Blood, vol. 115, no. 18, pp. 3664–3670, 2010, doi: https://doi.org/10.1182/blood-2009-07-231324.