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Case Report

Acute Abdominal Presentation of Enteropathy Intestinal T-cell Lymphoma: A Rare Case Report

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Abstract

Intra-abdominal hollow organ perforation is a critical condition that requires immediate intervention. There are various causes of small bowel perforation, including benign diseases, iatrogenic procedures, and perforation associated with tumors. However, preoperative imaging alone is often insufficient to determine the underlying cause. In this report, we present a unique case involving a malignant neoplasm of the small intestine resulting in bowel perforation.

Keywords: Hollow organ perforation, monomorphic epitheliotropic intestinal T-cell lymphoma, non-Hodgkin lymphoma

Introduction

Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) is a rare, aggressive gastrointestinal tract lymphoproliferative disorder accounting for <5% of all gastrointestinal tract lymphomas. The symptoms of MEITL include abdominal pain, bowel perforation, diarrhea, bowel obstruction, and gastrointestinal bleeding. The prognosis is poor, and standard treatment involves surgery, chemotherapy, and autologous stem cell transplantation (ASCT). Due to its rarity and aggressive nature, further research and awareness are imperative for improving outcomes for individuals affected by MEITL.

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Case Report

A 47-year-old woman presented with fever and abdominal pain lasting for 1 day. A physical examination revealed tenderness throughout her lower abdomen along with rebounding pain. Laboratory test results indicated leukocytosis (9.44×10^3/L white blood cell count, 83.8% neutrophils) and an elevated C-reactive protein level (13.4 mg/dL). Abdominal computed tomography (CT) revealed pneumoperitoneum and thickening of the small intestine wall [Figure 1]. Due to suspected small bowel perforation, an emergency exploratory laparotomy was scheduled.

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During surgery, a 7-cm tumor lesion was discovered in the ileum [Figure 2], invading the bowel wall. Tumor necrosis was observed, leading to bowel perforation. Another tumor measuring 4 cm was found in the upper jejunum. Fortunately, no peritoneal tumor metastasis was detected. The surgical team successfully performed segmental resection of the small intestine, along with mesenteric lymph node dissection.

The pathological examination findings were diffuse infiltration of small monotonous lymphoid cells from the mucosal surface into the regional adipose tissue and serosal layer. The Nuclear atypia of tumor cell was mild. Immunohistochemical tests showed that the tumor cells were diffusely positive for tumor marker CD3 and CD56, and negative for CD20, CD30, CD21, and CD5, primarily positive for CD8 (CD8:CD4 = 3~4:1). The Ki-67/MIB-1 index was approximately 60%–70%, and EBV-encoding RNA (EBER) was negative. These findings indicated primary malignant T-cell lymphoma, specifically monomorphic epitheliotropic type (MEITL).

Following a smooth recovery with restored bowel function and normal daily activities, the patient was discharged on day 30 postsurgery. She then received six cycles of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) chemotherapy. Following this initial course, a positron emission tomography-CT scan was conducted to assess the treatment response. The results indicated local recurrence of the intra-abdominal tumor, although there were no signs of bowel obstruction. As a salvage therapy, she received the etoposide, cisplatin, high-dose Ara-C, and methylprednisolone regimen followed by stem cell harvest. Subsequently, she received nine cycles of chemotherapy with Folotyn. Due to progressive disease, she received two cycles of dexamethasone, cisplatin, and high-dose Ara-C and one cycle of gemcitabine, dexamethasone, and cisplatin. She then received bendamustine, etoposide, cytarabine, and melphalan conditioning with autologous peripheral blood stem cell transplantation.

One month later, the patient suffered an intra-abdominal infection. As abdominal CT revealed an intra-abdominal abscess, explorative laparotomy was performed. The findings showed tumor recurrence with small intestine perforation at the distal jejunum, and small bowel resection and abscess drainage were performed. However, due to refractory intra-abdominal infection and septic shock, she died on post operation day (POD) 28. The total survival course after the initial diagnosis was 15 months.

Discussion

MEITL is an aggressive disease involving the small intestine, and symptoms of bowel obstruction or perforation are common.[1] MEITL is associated with deleterious mutation(s) and/or deletion of the *SETD2* gene, and 90% of cases have defective H3K36 trimethylation. Other frequently mutated genes in MEITL include *STAT5B* (57%), *JAK3 (*50%), *TP53 (*35%), *JAK1 (*12.5%), *BCOR,* and *ATM* (11%).[2] MEITL has no definite association with celiac disease, and it is more prevalent in Asian populations. The most commonly involved site is the jejunum.[3] The median overall survival of MEITL is 10 months, with the majority of cases presenting at stage I or II. The median duration of symptoms before diagnosis is 2 months. Treatment typically involves surgery and chemotherapy, followed by ASCT.[4] An anthracycline-based CHOP regimen is commonly used, with the addition of brentuximab vedotin in patients diagnosed as CD30 positive. Haddad and Malireddy reported that the combination of chemotherapy and stem cell transplant for MEITL demonstrated statistically superior outcomes compared to no treatment, with median overall survival times of 11, 22, and 0.75 months, respectively.[5] Further analysis revealed that surgical resection, in combination with chemotherapy and ASCT, conferred a survival advantage. In addition, combination treatments with anthracyclines and etoposide showed better overall survival, and the quality of response to treatment impacted the outcomes. Yi *et al*. reported that factors associated with more favorable outcomes included younger age, better performance status, earlier Lugano stage, chemotherapy, achieving complete remission, and receiving ASCT.[6] They also reported that the relatively short progression-free survival of 6.9 months in their study

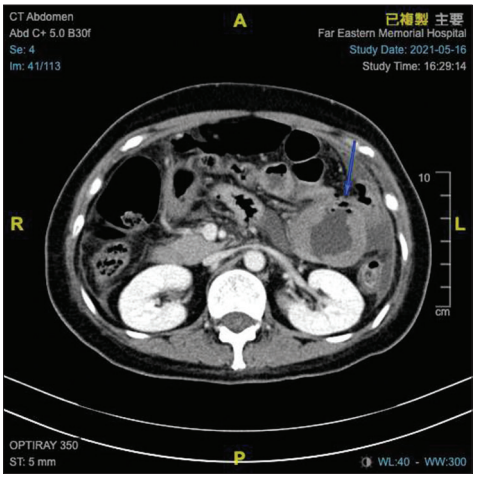

Figure 1: Pneumoperitoneum and thickening of the small intestine wall

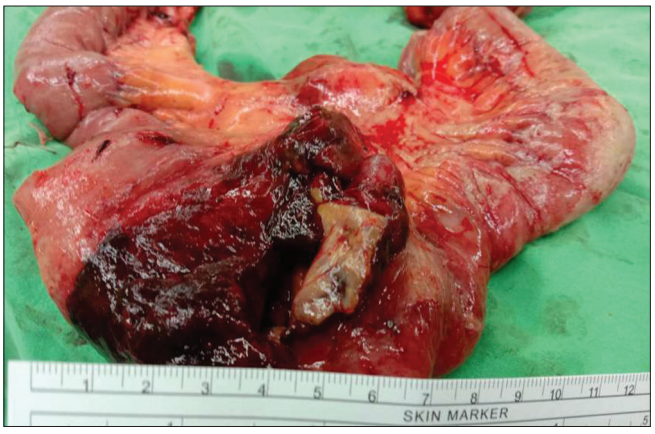

Figure 2: Ileum tumor

likely contributed to the high rate of relapse observed in the gastrointestinal tract, where 96% of documented relapses occurred.

In summary, MEITL is a rare and aggressive gastrointestinal lymphoproliferative disorder. Early diagnosis and appropriate treatment, including surgery, chemotherapy, and stem cell transplantation, are crucial for improving the outcomes of MEITL patients. Further research is needed to better understand the heterogeneity of this disease and develop targeted therapies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Data availability statement

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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