Case report and literature review of monomorphic epitheliotropic intestinal T-cell lymphoma

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ABSTRACT

Background: Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) is a rare, highly aggressive primary intestinal T-cell lymphoma predominantly observed in male populations in Asia and Latin America. The disease most commonly involves the proximal small intestine, particularly the jejunum, and is characterized by non-specific clinical manifestations.

Case Presentation: We report the case of a 37-year-old male who presented with periumbilical abdominal pain of indeterminate etiology, which subsequently migrated to the left upper quadrant. Computed tomography of the abdomen revealed focal wall thickening in the small intestine within the right lower quadrant and left middle-upper abdomen, alongside marked irregular thickening and rigidity in the left upper abdomen, findings suggestive of a neoplastic lesion. Intraoperatively, a firm mass measuring approximately 8 cm × 10 cm was identified in the small intestine. Histopathological examination confirmed the diagnosis of MEITL; notably, the tumor cells exhibited expression of the B-cell immunohistochemical marker CD20.

Conclusion: MEITL is characterized by a low incidence and poses significant challenges for early detection. T-cell lymphomas co-expressing CD20 are rare, and the prognostic implications of CD20 expression in MEITL remain controversial. Further studies with larger sample sizes are warranted to elucidate this relationship, to facilitate more precise treatment strategies, and to improve patient outcomes.

Keywords: Intestinal T-cell lymphoma, monomorphic epitheliotropic intestinal T-cell lymphoma, immunohistochemistry, prognosis.

Type of Article: Case Report **Specialty:** Pathology

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Background

Intestinal T-cell lymphoma is a rare neoplasm, accounting for 0.25% of all malignant lymphomas and less than 5% of primary gastrointestinal tract lymphomas. It originates from the malignant proliferation of intraepithelial lymphocytes and is classified into distinct subtypes, including enteropathy-associated T-cell lymphoma (EATL), monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL), indolent T-cell lymphoproliferative disease of the gastrointestinal tract, and intestinal T-cell lymphoma, not otherwise specified [1]. MEITL, formerly designated as type II EATL, represents a rare extranodal lymphoma arising from intraepithelial T lymphocytes.

This case report describes a young patient presenting with abdominal pain who was diagnosed with MEITL following small intestinal resection, with concurrent involvement of the colon and perimesenteric lymph nodes. Immunohistochemical analysis revealed expression of the B-cell marker CD20, the prognostic significance of which remains controversial. The patient has been followed up for 12 months and remains in good clinical condition. This report delineates the typical histomorphological features and immunohistochemical profile of MEITL and discusses prognostic factors, aiming to facilitate early detection and diagnosis to optimize clinical management and patient outcomes.

Case Report

In March 2025, a 37-year-old male presented with periumbilical pain of insidious onset, characterized by paroxysmal colic lasting several minutes per episode with spontaneous remission. Subsequently, the symptomatology migrated to the left upper quadrant, evolving into postprandial distension and discomfort. Laboratory

investigations, including complete blood count, biochemical panels, and tumor markers, yielded results within normal limits. Contrast-enhanced computed tomography (CT) of the abdomen revealed localized small intestinal wall thickening in the right lower quadrant and left middle upper abdomen. Notably, the intestinal wall in the left upper abdomen exhibited marked irregular thickening and rigidity with significant contrast enhancement. Multiple enlarged lymph nodes with homogeneous enhancement were identified in the adjacent mesentery, alongside peritoneal thickening and blurring in the right upper abdomen. These radiographic findings were suggestive of a neoplastic lesion, with suspected lymph node metastasis and peritoneal seeding. Intraoperative exploration revealed a moderate amount of ascites. A hard, irregular mass measuring approximately 8 cm × 10 cm was identified within the small intestine; the mass was densely adhered to the greater omentum and the hepatic flexure of the transverse colon, with evidence of local infiltration.

Pathological Examination

Microscopic Findings: Low-power examination revealed transmural lymphoid infiltration, accompanied by necrosis on both the serosal and mucosal surfaces. Under high-power magnification, the neoplastic cells appeared monomorphic, characterized by round or polygonal, hyperchromatic nuclei with folded and indented nuclear membranes, coarsely granular chromatin, inconspicuous nucleoli, and pale to eosinophilic cytoplasm. Cellular atypia was not pronounced. Villous atrophy was observed adjacent to the ulcer, without associated crypt hyperplasia, and prominent epitheliotropism was identified Figure 1.

Immunohistochemical Findings: The tumor cells expressed CD2, CD3, CD8, CD7, CD56, lysozyme, TIA-1, Bcl-2, and CD20, with focal CD4 expression. AE1/AE3 immunostaining highlighted the epitheliotropism. The Ki-67 proliferation index was approximately 60%. The neoplastic cells were negative for Bcl-6, CD10, ALK, CD21, PAX-5, CD79α, CD5, and CD30. In situ hybridization for Epstein–Barr virus-encoded small RNAs (EBER) yielded negative results Figure 2.

Pathological diagnosis

Based on morphological and immunohistochemical findings, the diagnosis supports T-cell lymphoma, consistent with monomorphic epitheliotropic intestinal T-cell lymphoma; tumor cells penetrate the serosa, with tumor involvement observed in the greater omentum; no definitive tumor metastasis is identified in perienteric lymph nodes (0/14); tumor involvement is not observed at the resection margins of the submitted colon and small intestine. According to the Lugano staging system for primary gastrointestinal lymphoma, this case is classified as Stage IIE.

Follow-up

The patient was discharged after surgery and did not receive further treatment at our hospital. The patient independently sought consultation and treatment at a higher level hospital (the patient reported undergoing chemotherapy, but specific details are unknown). A follow-up CT scan performed at the higher level hospital three months after treatment revealed no enlarged lymph nodes. Upon recent follow-up with the patient's family (April 7, 2026), the patient reported being in good condition and having achieved partial remission.

Discussion

The association between intestinal lymphoma and celiac disease was first reported by Gough et al. in 1962 [2]. Subsequently, Chott et al. described a distinct lymphoma characterized by a monomorphic proliferation of small-to-medium-sized T cells, designating it as CD56+ intestinal T-cell lymphoma [3]. In the revised 4th edition of the WHO classification of Tumours of Haematopoietic and Lymphoid Tissues (2017), type I EATL, which is associated with celiac disease, retained its nomenclature, whereas type II EATL, unrelated to celiac disease, was reclassified as MEITL [4]. The 2022 WHO classification further recognized MEITL as a distinct and rare subtype of primary intestinal T-cell lymphoma, distinguished by its unique pathological and epidemiological features [5].

Clinically, MEITL is an uncommon, highly aggressive neoplasm that predominantly affects males in Asia and Latin America. It most frequently arises in the proximal small intestine, particularly the jejunum. Clinical manifestations are often nonspecific and may include abdominal pain, obstruction, perforation, and weight loss [6]. Metastasis to sites, such as the central nervous system, spleen, liver, and gallbladder, has been documented [7], and the median survival time is approximately 7 months [8,9]. The differential diagnosis of MEITL typically includes: 1. Enteropathy-associated T-cell lymphoma (EATL): The tumor cells are predominantly medium-to-large in size, necrosis is common, and an inflammatory background is frequently present; on the contrary, MEITL cells are monomorphic, necrosis is rare, and the inflammatory background is inconspicuous. Immunohistochemically, the tumor cells express CD3, CD7, CD8, and TIA-1, but do not express CD5 or CD4, and are negative for CD56. 2. Indolent T-cell lymphoma of the gastrointestinal tract: Tumor cell infiltration is superficial, without full-thickness invasion of the intestinal wall, and epitheliotropism is rare. Immunohistochemically, the tumor cells express CD2, CD3, CD5, CD7, and CD8, but do not express CD4 or CD56, and the Ki-67 proliferation index is low (<10%). 3. Extranodal NK/T-cell lymphoma, nasal type: Tumor cells often exhibit angioinvasion, geographic necrosis is common, cell size varies, and nuclear atypia is distinct. Immunohistochemically, the tumor cells express CD3

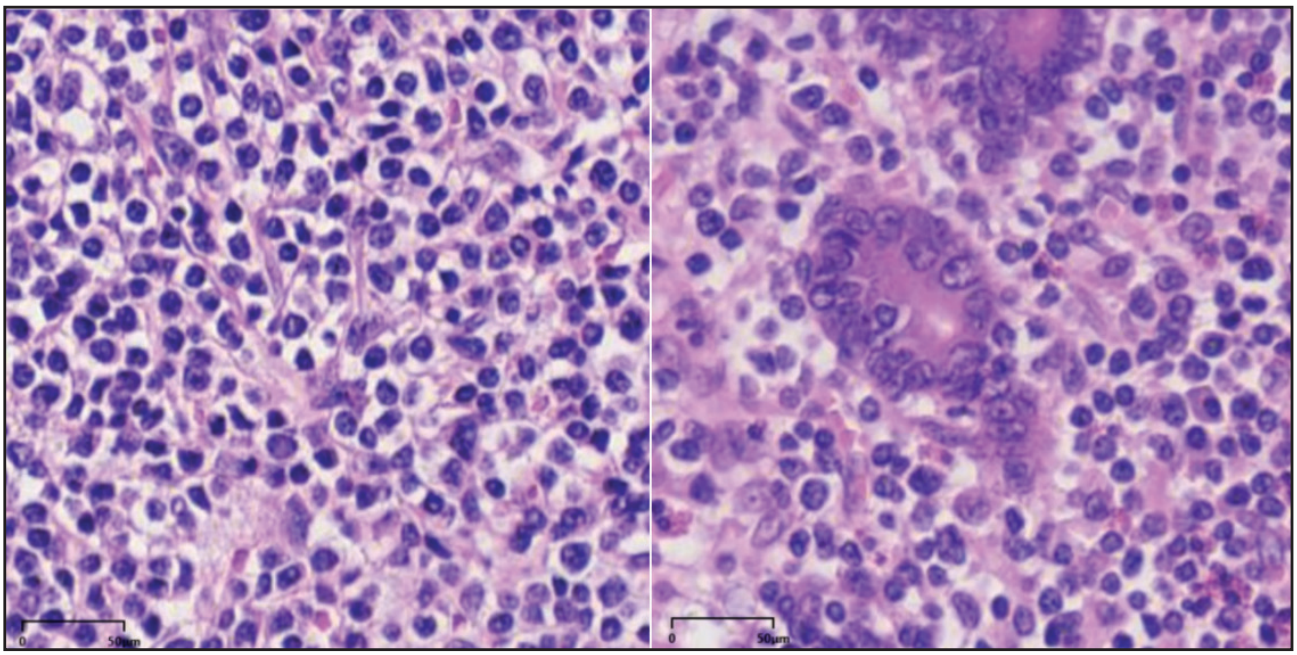

Figure 1. *The tumor cells exhibit uniform morphology, with round or polygonal nuclei and hyperchromasia, showing obvious epitheliotropism.*

(cytoplasmic type) and CD56, do not express CD5, and are positive for EBER. 4. Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma): Tumor cells exhibit variable morphology, resembling follicular center cells, small lymphocytes, or monocytoid B cells, and are often accompanied by plasma cell differentiation. Immunohistochemically, they express B-cell markers such as CD20 and CD79a, do not express T-cell markers, and have a low Ki-67 proliferation index. 5. Poorly differentiated carcinoma: Tumor cells exhibit an epithelioid appearance, arranged in nest-like or gland-like structures, often accompanied by necrosis and vascular invasion. Immunohistochemically, the tumor cells express CK and are negative for EBER.

In the present case, the lesions were predominantly localized to the small intestine, characterized by an insidious onset, atypical clinical manifestations, and recurrent abdominal pain. Microscopically, MEITL is defined by monomorphic small-to-medium-sized tumor cells and the presence of lymphoepithelial lesions. In small intestinal involvement, the villous structures are typically distorted and expanded; concomitant with an increase in intraepithelial lymphocytes, tumor cells frequently infiltrate the surrounding mucosa while the villous architecture remains relatively preserved. Immunohistochemically, tumor cells typically exhibit positivity for CD2, CD3, CD7, CD8, CD56, and TIA-1, alongside a high Ki-67 proliferation index, but are negative for CD4, CD5, and CD30, with rare exceptions [10]; these findings are consistent with the present case. Notably, the tumor cells in this instance demonstrated aberrant CD20 expression. Studies have indicated that CD20-positive T-cell lymphomas are relatively rare. Two retrospective analyses [11,12] have indicated that CD20 expression may signify disease relapse or progression and is associated with a relatively poor prognosis.

On the contrary, a 2023 study by international scholars [13] involving 71 patients with MEITL conducted a comprehensive clinical, pathological, and genomic analysis, revealing that CD20 expression (observed in 20% of cases) correlated with a favorable prognosis. These findings contradict previous reports, a discrepancy likely attributable to limited sample sizes; consequently, the prognostic significance of B-cell marker expression in MEITL warrants further validation through studies employing larger cohorts.

In a recent retrospective analysis of 32 cases of MEITL in South China, Guo et al. [14] demonstrated that MEITL exhibits distinct clinicopathological features compared to other subtypes, such as ENKTL, and other intestinal T-cell lymphomas. High expression of spleen tyrosine kinase (SYK) and PD-L1 was identified as a significant predictor of poor prognosis ($p < 0.001$). Furthermore, the authors proposed that SYK may serve as a valuable diagnostic marker for MEITL, particularly in cases negative for CD8 or CD56; these findings align with the observations of Mutzbauer et al. [15]. The overall prognosis for MEITL remains poor, with diagnostic delay being a critical determinant of adverse outcomes. A multicenter retrospective study of 42 patients [16] indicated that younger age, favorable performance status, early-stage disease (Lugano classification), and autologous stem cell transplantation (ASCT) are associated with improved prognosis.

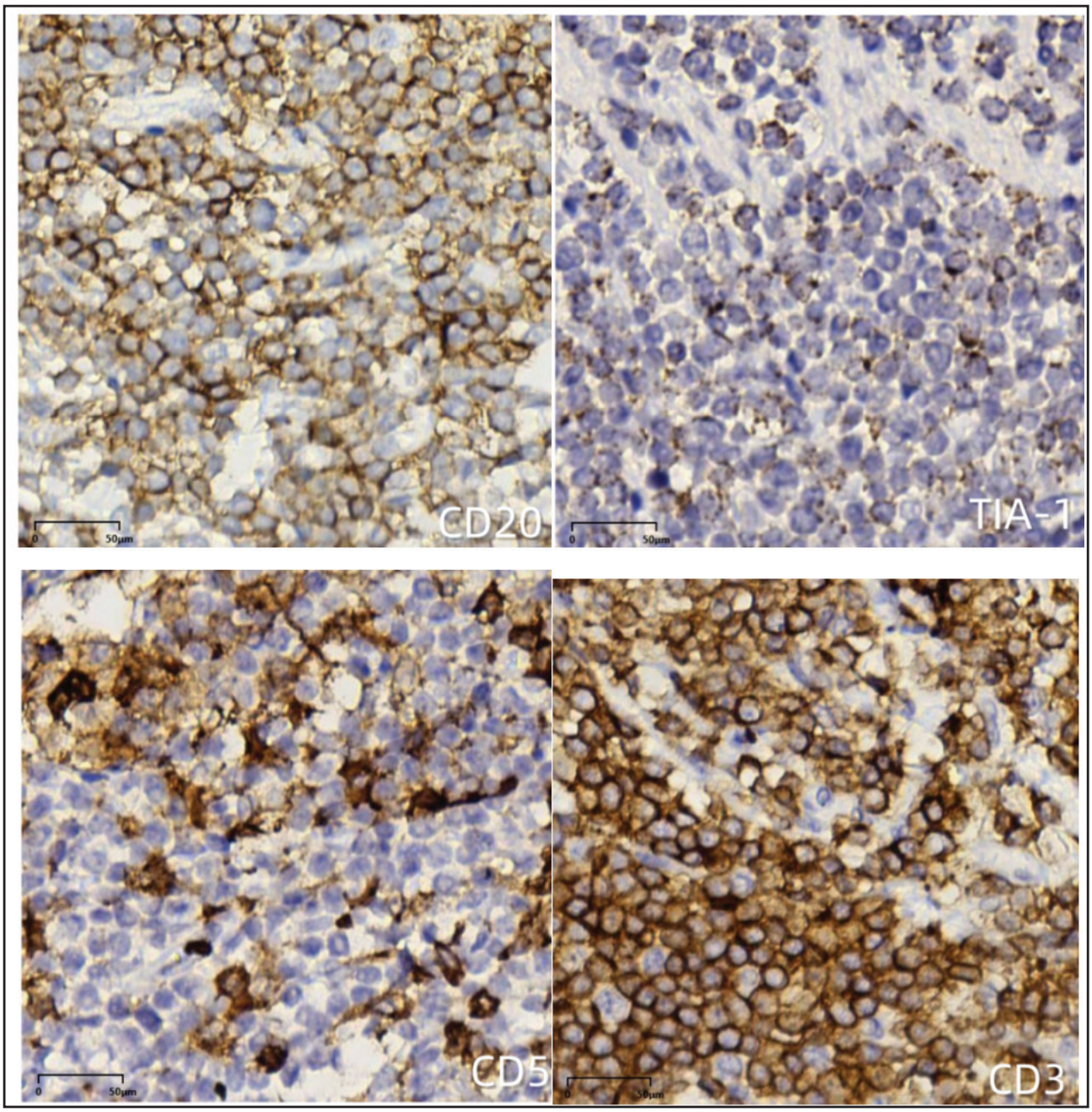

Figure 2. *Immunohistochemistry (En Vision method). Tumor cells are CD20 positive (moderate intensity); tumor cells are positive for TIA; tumor cells are negative for CD5; tumor cells are positive for CD3.*

Currently, no standardized treatment regimen has been established for MEITL. While the CHOP regimen (cyclophosphamide, doxorubicin, vincristine, and prednisone) is widely utilized for T-cell lymphomas, its prolonged administration is associated with significant cardiotoxicity, including congestive heart failure and chronic irreversible cardiac damage. Conversely, pegylated liposomal doxorubicin (PLD) has been demonstrated to significantly mitigate cardiotoxicity while preserving antitumor efficacy. Notably, a case study reported a patient with pathologically confirmed MEITL, who survived for 1 year without cardiotoxicity following treatment with a CHOPE regimen incorporating PLD [17]. Regarding novel therapeutic approaches, Timothy et al. [18] reported that chimeric antigen receptor T-cell (CAR-T) therapy induced durable remission in patients with CD30-positive enteropathy-associated T-cell lymphoma (EATL). Furthermore, a retrospective analysis of 35 MEITL patients by Korean researchers [19] suggested that ifosfamide-based chemotherapy followed by allogeneic hematopoietic stem cell transplantation (allo-HSCT) may improve prognosis, implying that chemotherapy alone is often insufficient for sustaining long-term remission; notably, two subjects within this cohort achieved complete remission following allo-HSCT alone. In addition, an international case

report described a 68-year-old female MEITL patient who achieved complete remission and remained recurrence-free for 5 years after completing eight cycles of CHOP, suggesting that early and effective intervention may facilitate long-term survival [20].

In summary, we present a case of a young patient with MEITL primarily located in the small intestine, with involvement of the colon and mesenteric lymph nodes, exhibiting tumor cell expression of B-cell markers. The patient remains in good clinical condition following a 12-month follow-up period. Given the low incidence and diagnostic challenges associated with MEITL, as well as the rarity of T-cell lymphomas co-expressing CD20, the prognostic implications of B-cell marker expression remain controversial. Consequently, future studies with larger sample sizes are warranted to elucidate these characteristics, with the goal of optimizing therapeutic strategies and improving patient prognosis.

Conclusion

We report a case of a young patient with MEITL involving the small intestine, colon, and perimesenteric lymph nodes, whose tumor cells aberrantly expressed the B-cell marker CD20. The patient remains in favorable clinical condition after 12 months of follow-up. MEITL is a rare and highly aggressive lymphoma with a low incidence, posing significant challenges for early diagnosis. Co-expression of CD20 in T-cell lymphomas is exceptionally rare, and its prognostic significance in MEITL remains controversial, with conflicting results from limited studies. Therefore, larger-scale investigations are warranted to clarify the role of B-cell marker expression, in order to refine risk stratification and guide more effective therapeutic strategies for this aggressive disease.

List of Abbreviations

allo-HSCT	Allogeneic hematopoietic stem cell transplantation
ASCT	Autologous stem cell transplantation
CAR-T	Chimeric antigen receptor T-cell
CT	Computed tomography
CHOP	Cyclophosphamide, doxorubicin, vincristine, prednisone
EATL	Enteropathy-associated T-cell lymphoma
EBER	Epstein-Barr virus-encoded small RNAs
MEITL	Monomorphic epitheliotropic intestinal T-cell lymphoma
PLD	Pegylated liposomal doxorubicin
SYK	Spleen tyrosine kinase

Conflict of interest

The authors declare that they have no conflicts of interest regarding the publication of this case report.

Funding

None.

Consent for publications

The electronic informed consent form has been uploaded as an image in the attachment.

Ethical approval

Ethical approval is not required at our institution for anonymous case reports.

Patient consent

Written informed consent was obtained from the patient.

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Summary of the case

1	Patient (gender, age)	Male, 37 years old
2	Final Diagnosis	Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL)
3	Symptoms	Recurrent abdominal pain (periumbilical, later migrating to left upper quadrant) and postprandial abdominal distension
4	Medications	None (no adjuvant chemotherapy or targeted therapy administered)
5	Clinical Procedure	Exploratory laparotomy with segmental resection of the small intestine and adherent colon, plus regional lymph node dissection
6	Specialty	Pathology, Gastroenterology, Surgical Oncology