

Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL)

– A case report

Kevin KC Cheung, Alex KH Tsang

Yan Chai Hospital

Introductions

Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL), formerly known as type 2 enteropathic-associated T-cell lymphoma, is a rare type of peripheral T-cell lymphoma that is not associated with coeliac disease and is relatively more common in Asian and Hispanic patients. On the other hand, the former type I enteropathic-associated T-cell lymphoma (EATL), which is now designated simply as EATL, is typically associated with coeliac disease. The two types of intestinal T-cell lymphoma show distinct clinical, histological and immunological features. We report a case of MEITL in a 66-year-old gentleman, highlighting the importance of differentiating from other intestinal T cell lymphomas.

Case Report

Clinical History

The patient was a 66-year-old Chinese male with hypertension and diabetes who presented with abdominal distension and weight loss for one year. There was no history of chronic malabsorption. Computed Tomography scan revealed small bowels with mural thickening and focal dilatation, suggestive of aggressive tumour with suspected concealed rupture. Suspected nodal and lung metastases were also identified on imaging. Emergency operation was performed and loops of adhering small intestine including a sizeable small intestine tumour with bleeding and rupture was identified.

Macroscopic Examination

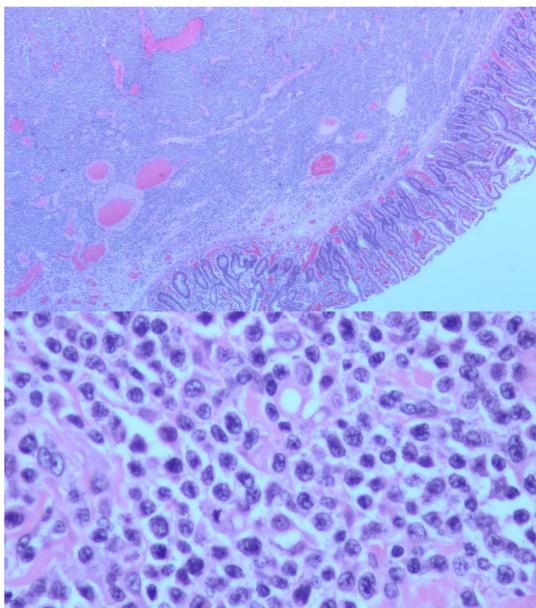
The resected small intestine consisted of three loops of small intestine adhering to one another. There was perforation of intestine wall with a 4cm necrotic tumour between the three loops of small intestine. Cut section at the perforation sites showed presence of pinkish soft tumour tissue.

Microscopic appearance

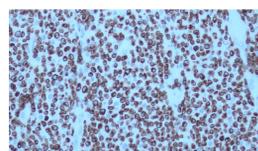
It showed small intestinal wall with diffuse sheet-like infiltrates of monotonous medium-sized lymphoid cells associated with perforation and peritonitis. The lymphoid cells exhibited folded nuclei, indistinct nucleoli and scanty cytoplasm. Mitotic and apoptotic figures were seen. Epitheliotropism is focally seen. One lymph node is noted and there is lymphoma involvement.

Immunohistochemical study

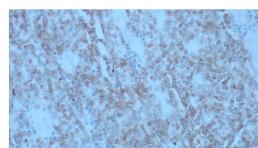
The lymphoid cells are diffusely positive for T-cell markers CD3, CD8 and CD56. They are negative for CD5 and CD4. B-cell marker (CD20), CD10, TdT and EBER are negative. They are negative for TCRgamma or TCRbeta (T-cell receptor-silent).



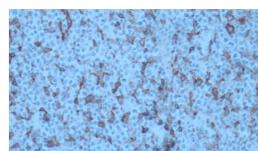
Low and high power view showing sheet like medium-sized lymphoid infiltrates.



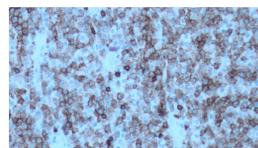
CD3



CD56



CD4



CD8

Discussions

In a multicenter study, the most common presenting features among a study group of 38 patients within a 19-year period were perforation (34%), pain (32%), and obstruction (21%). Radiology can help identify the presence of tumour but the specific diagnosis requires histological examination.

The typical histological features of the neoplastic lymphocytes are described to be medium in size with monomorphic nuclei and a rim of pale cytoplasm. The nuclei show finely dispersed chromatin and inconspicuous nucleoli. Epitheliotropism is common.

MEITL has a distinctive immunophenotype. The neoplastic lymphocytes are typically positive for CD3, CD8, and CD56. Most tumours lack CD5. Expression of T-cell receptor (TR) gamma is often positive, although some cases express TR beta or are TR-silent. Rarely both TR gamma and TR beta are expressed. TIA1 is usually positive, but expression of other cytotoxic molecules (including granzyme B and perforin) is less consistent. Approximately 20% of cases show aberrant expression of CD20.

The differential diagnoses of MEITL include EATL, intestinal NK/T-cell lymphoma, and indolent T-cell lymphoproliferative disease. EATL generally has a polymorphic cellular composition and negative for CD8 and CD56. Staining with MATH can also help differentiate MEITL over EATL. NK/T-cell lymphoma commonly shows angiocentric/angiodestructive growth and necrosis. EBER is helpful in differentiating NK/T-cell lymphoma from MEITL, as it is positive in the former and negative in the latter. As for indolent T-cell lymphoproliferative disorder the clinical course is chronic and indolent and the neoplastic lymphocytes are non-destructive small lymphocytes negative for CD8 and CD56.

Clonal rearrangement of the TR genes is seen in > 90% of cases. Common genetic alterations include extra signals for MYC and gains at 9q34. Activating mutations in STAT5B have also been identified in a high proportion of cases. JAK3 and GNAI2 are also mutated in some cases

MEITL is an aggressive form of T-cell lymphoma with poor prognosis. The median survival is 7 months. No specific guideline is available for the treatment of MEITL. Treatment includes surgery, high-dose chemotherapy, and autologous stem cell transplantation.

Conclusions

We report a case of MEITL that presented as perforation of small intestine, weight loss and abdominal discomfort. It should be considered in patients with atypical lymphoid proliferation in the small intestine. Appropriate immunostaining and molecular studies should be applied to differentiate from other differential diagnoses.

References

1. Tse E, Gill H, Loong F, Kim SJ, Ng SB, Tang T. Type II enteropathy-associated T-cell lymphoma: A multicenter analysis from the Asia lymphoma study group. *Am J Hematol* 2012;87:663-8
2. Kim YS, Choi YS, Park JS, Kim BG, Cha SJ, Chi KC, et al. Case of small bowel perforation due to enteropathy-Type T-cell lymphoma. *Yonsei Med J* 2009;50:859-61.
3. Jaffe ES, Chott A, Ott G, Chan JK, Bhagat G, Tan SY, et al. Monomorphic epitheliotropic intestinal T-cell lymphoma. In: Swerdlow SH, Campo E, Harris NL, Jaffe ES, Pileri SA, Stein H, editors. *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissue*. 4th ed. Lyon: IARC Press; 2017. p. 377-9