OPEN ACCESS – CASE REPORT

Monomorphic Epitheliotropic Intestinal T-cell Lymphoma with Alveolar and Maxillary Metastases: A Case Report

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ABSTRACT

This is a case of monomorphic epitheliotropic intestinal T cell lymphoma presenting as ruptured viscous in a 64-year-old male. Monotonous cells were seen to infiltrate the full thickness of the bowel with immunoreactivity for CD3, CD8, CD56, CD68 and Granzyme B. The patient subsequently presented with maxillary and alveolar mass with similar immunohistomorphologic findings, suggesting metastasis. This rare disease has poor prognosis thus further reports should be conducted to profile this type of tumor.

Key words: monomorphic epitheliotropic intestinal T-cell lymphoma; intestinal perforation; enteropathy-associated T-cell lymphoma type II; case report

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INTRODUCTION

Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) is a rare and aggressive type of tumor derived from intraepithelial T lymphocytes. This entity was previously grouped under Type II Enteropathy associated T-cell lymphoma but was re-classified as a distinct entity due to its lack of association with celiac disease.[1] The disease has a worldwide distribution, however, most reports are seen in the Asian population, with most cases occurring in East Asia.[2-5] MEITL and EATL comprise 2% of known cases of peripheral T-cell lymphomas in Asia. In the Philippines, cases of primary intestinal T-cell lymphoma are underreported. As of writing, there is no known case of MEITL reported in recent literature locally.

CASE

We report a case of a 64-year-old male who presented in the emergency room with a 3-day history of sharp periumbilical pain which eventually worsened and became associated with fever. Patient has no known comorbidities, or history of celiac disease. He is a 20-pack year smoker, with a family history of emphysema. Physical examination findings showed stable vital signs but with distended abdomen, direct and rebound tenderness on all quadrants with guarding. Chest X-ray revealed pneumoperitoneum thus a ruptured viscus was considered.

The patient was subsequently admitted and underwent an exploratory laparotomy where intraoperative findings showed a necrotic segment of jejunum 10 to 20 cm from the Ligament of Treitz (LOT), thick walled and perforated ileum located 200 cm from LOT, and multiple palpable mesenteric lymph nodes. Ileal resection, gastrojejunostomy, and double barrel ileostomy was done, and the specimen submitted for histopathology.

The patient was discharged well. Three weeks after the initial presentation, the patient noted enlarging right maxillary and left alveolar mass, warranting consultation in the outpatient department. Biopsy done revealed similar

microscopic and immunohistochemical findings with the intestinal specimen. One month after initial admission, the patient was readmitted for generalized abdominal pain, vomiting with noted hypotension, guarding and abdominal distention on physical examination. Patient was managed as a case of ruptured viscous and underwent re-exploratory laparotomy and partial duodenojejunectomy. During his hospital stay, the patient had episodes of hypotension, desaturation, and melena. The patient was referred to hospice and palliative care but eventually opted discharge and was lost to follow up.

Segments of ileum and jejunum were sent for histopathologic examination. Both ileal and jejunal resection specimens showed multiple, dark green, ill-defined masses which constrict but do not obstruct the lumen. The mass was also seen to grossly perforate the jejunum (Figure 1). On microscopy, monotonous small to medium round cells are seen to densely infiltrate the mucosa. The intestinal villi appear to be attenuated and are entirely lost in other areas (Figure 2). No glandular spacing or dropout is seen (Figure 3A). On high power view, the cells have vesicular nuclei and conspicuous nucleoli with scant cytoplasm (Figure 3B). Other areas of the intestine adjacent to the tumor show necrosis and ulceration (Figure 4A). Areas uninvolved by the tumor also show lymphoid infiltration, similar to lymphocytic colitis. (Figure 4B). The same microscopic features were also seen in the maxillary, alveolar and second resection specimens (Figure 5).

Immunohistochemistry studies showed positive expression for LCA, CD3, CD8, CD56, CD68 and Granzyme B (Figure 5). The tumor has negative staining for Pancytokeratin, CD20, CD30, CD4 and CD5 (Figure 6). Given the clinical and immunohistomorphologic findings, the diagnosis of Monomorphic Epitheliotrophic Intestinal T-cell Lymphoma was established.

DISCUSSION

MEITL is a primary intestinal T-cell lymphoma that is known to have a worldwide distribution but is mostly reported among the Asian and Hispanic population. Most reported cases are in the East Asian Region, but cases have also been reported in western countries, which share the same clinical course and histomorphologic findings.[6] Historically, MEITL was grouped under Enteropathy-type-T-cell lymphoma and designated as Type 2 Enteropathy associated T-cell lymphoma (EATL). In 2016, it was reclassified as a separate entity due to its lack of association

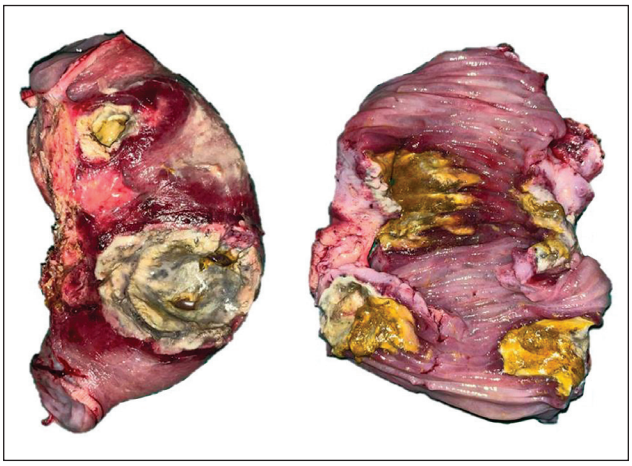

Figure 1. Resected segment of the ileum shows multiple ill-defined cream white to yellow necrotic lesions which perforate the full thickness of the intestinal wall

Photos courtesy of Dr. Darwin Del Rosario from the Surgery Department.

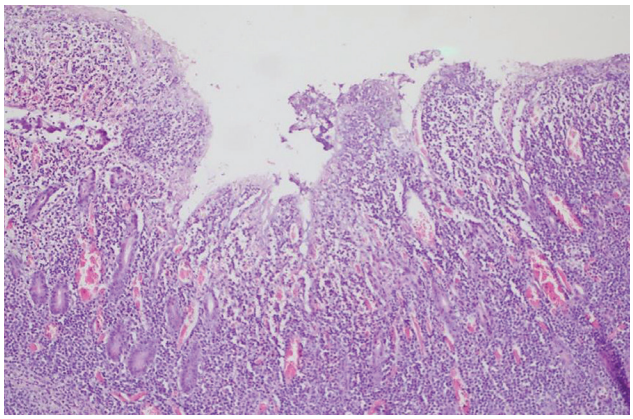

Figure 2. ***Intestinal Mass.*** Dense sheets of round blue cells are seen to infiltrate the mucosa, with attenuation of the small intestinal villi, and ulceration of the surface epithelium (H&E, 20x).

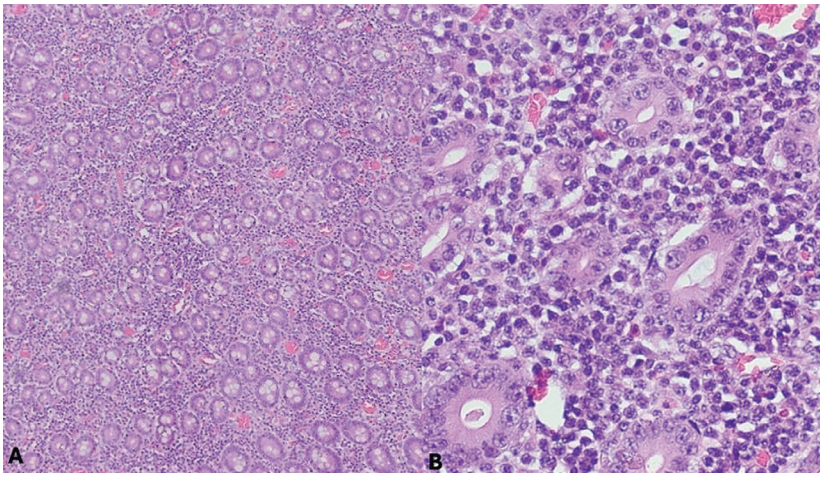

Figure 3. ***Intestinal Mass.*** Monomorphic round blue cells are seen to surround glands. **(A)** No increased glandular spacing or loss of glands are seen (H&E, 20x). **(B)** The cells are monomorphic with round to irregular vesicular nuclei, conspicuous nucleoli, and dense chromatin (H&E, 100x).

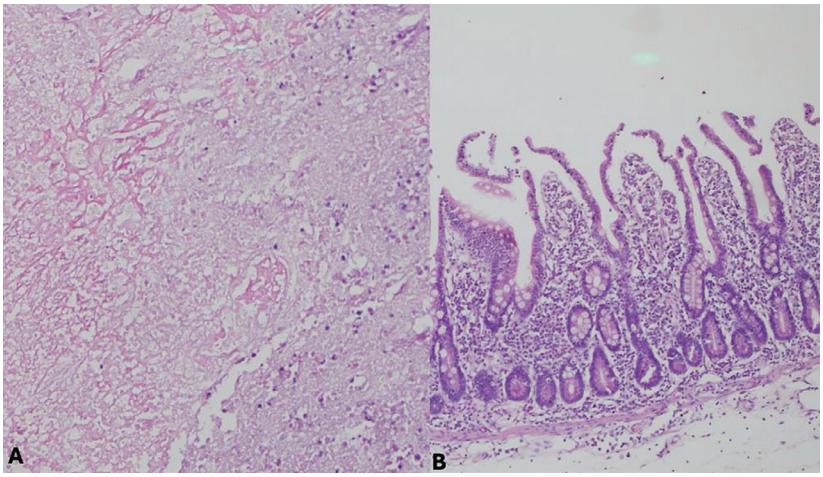

Figure 4. ***Intestinal Mass.*** **(A)** Other areas of the small intestine show necrosis (H&E, 20x). **(B)** Areas uninvolved by the tumor show normal architecture with intraepithelial lymphocytes (H&E, 20x).

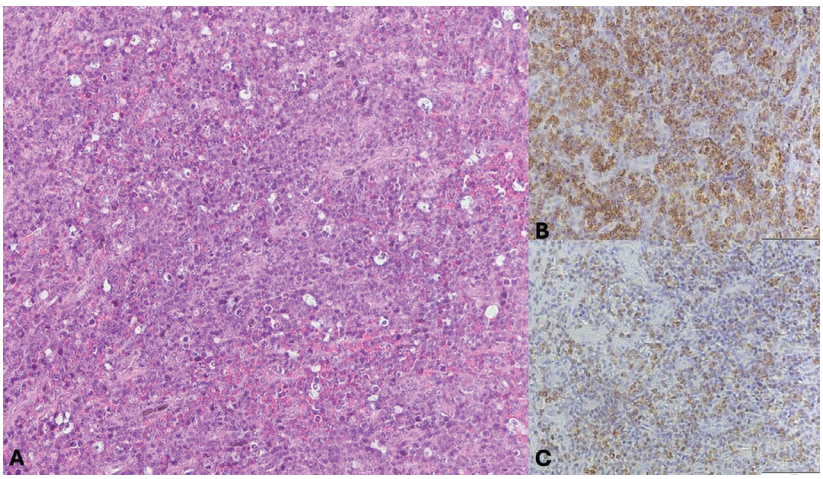

Figure 5. ***Alveolar Mass.*** **(A)** Morphologically similar monomorphic cells are also seen to infiltrate the alveolar mass, showing a "starry sky" appearance (H&E, 20x). **(B)** Tumor cells stained positive for CD3, confirming their T-cell lineage (HRP, 20x). C. CD20 only stained positively in the admixed non-neoplastic B cells (HRP, 20x).

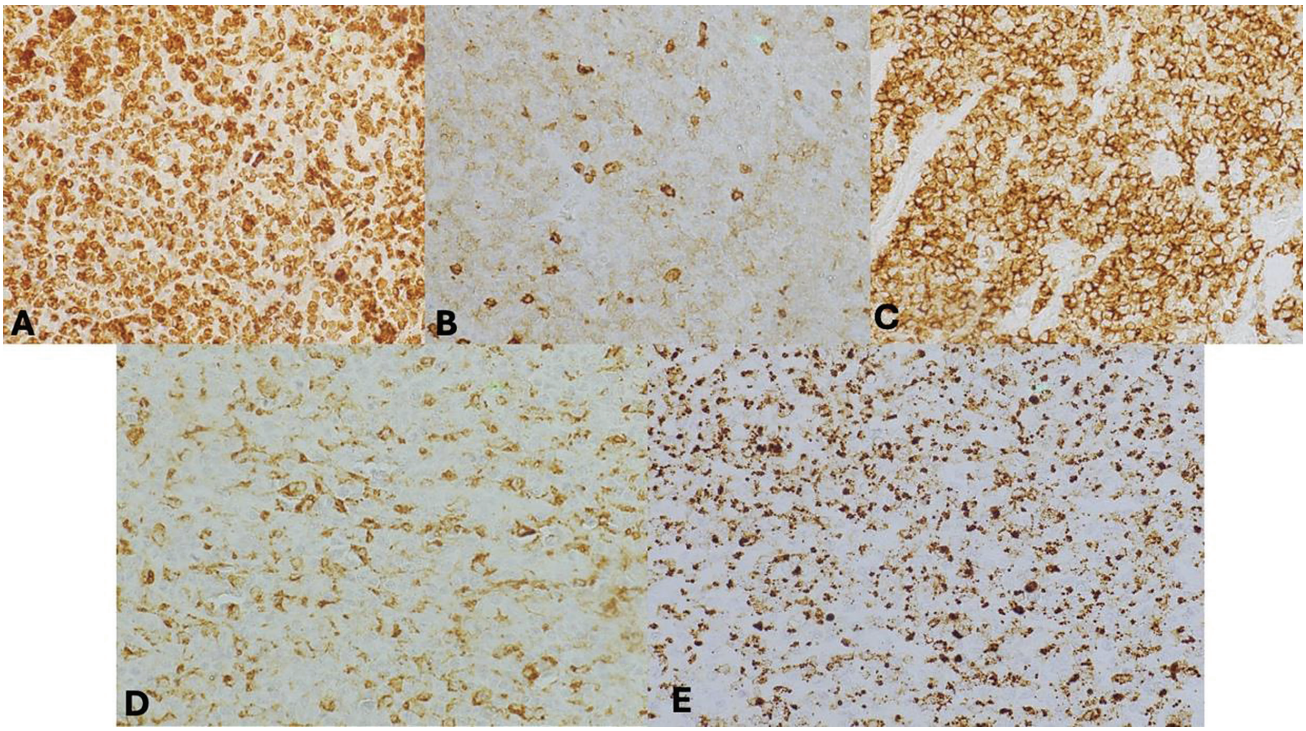

Figure 6. ***Immunohistochemistry profile of the intestinal mass.*** The mass showed: **(A)** Strong, diffuse positivity for CD3 (HRP, 40x); **(B)** Patchy positivity for CD8 (HRP, 40x); **(C)** Diffuse positivity for CD56 (HRP, 40x); **(D)** Diffuse positivity for CD68 (HRP, 40x); **(E)** Diffuse positivity for Granzyme B (HRP, 40x).

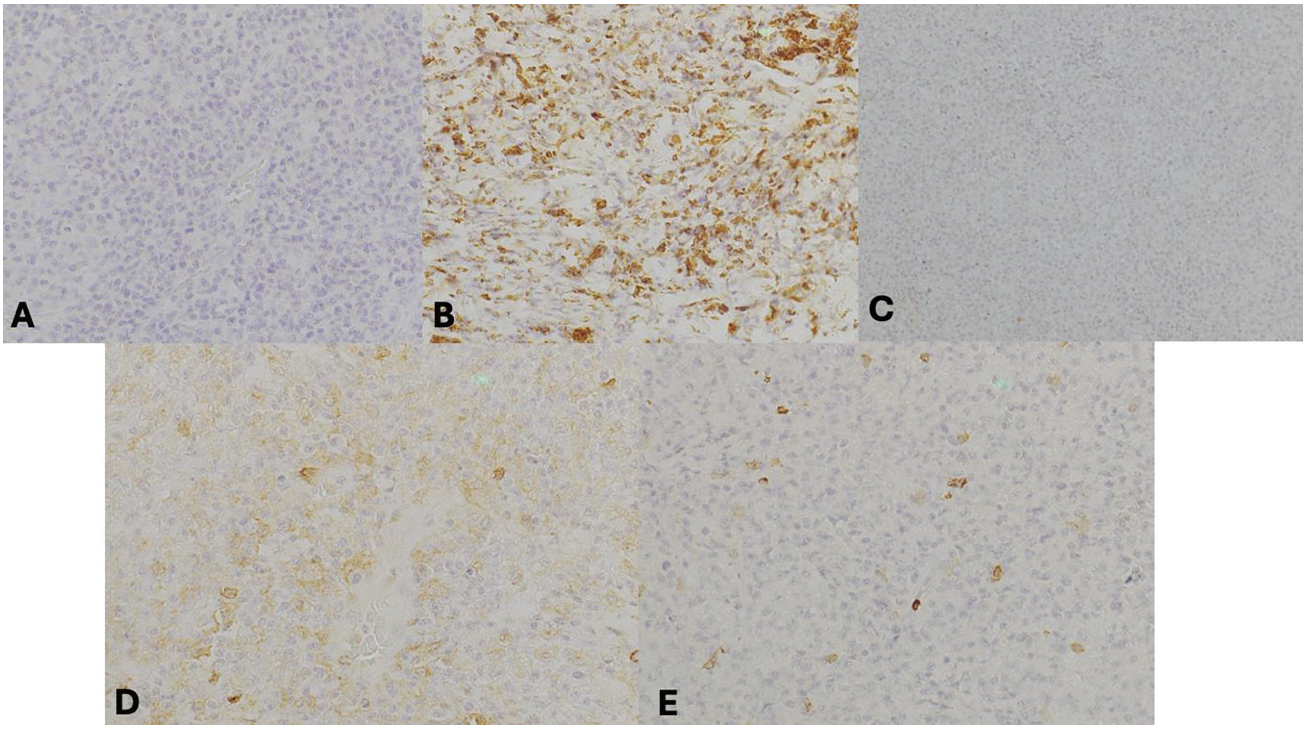

Figure 7. ***Immunohistochemistry profile of the intestinal mass.*** **(A-E)** The mass showed negativity for **(A)** Pancytokeratin, **(B)** CD20, **(C)** CD30, **(D)** CD4, and **(E)** CD5 (HRP, 40x).

with celiac disease, and its pathogenesis is less dependent on its presence. Genetic differences between the two entities supported this change in classification. MEITL showed MYC oncogene locus gain, and rare gains of chromosome 1q and 5q.[7] On the other hand, Type 1 Enteropathy associated T-cell lymphoma showed gains of chromosome 1q and 5q and HLA-DQB1 genotype patterns with celiac disease.

MEITL is reported to affect mostly adults, with an age range of 54-67 years old, with male predominance.[2] The disease usually presents with vague symptoms including abdominal pain, weight loss, and intestinal obstruction or perforation. MEITL most commonly affects the small intestine, with some cases being reported to affect the colon, stomach, and duodenum. The disease is known to disseminate to mesenteric lymph nodes, and in rare cases, in the lung, liver, and brain.[8-10] In the patient's case, the disease was noted to have metastasized to the bone, particularly in the right maxilla.

Because symptoms can be non-specific, patients tend to be diagnosed late and may not be able to start treatment early. The disease carries a poor prognosis, with only a median survival rate of 7 months, coupled with a low chemotherapy response rate on CHOP-like polychemotherapy. Despite this, cases with recurrence free survival for five years have been previously reported and those with complete surgical resection carry a better prognosis.[2,11]

Grossly, the tumor presents as an ulcerated mass and is indistinguishable from other hematolymphoid tumors. Microscopic examination shows monomorphic small round blue cells invading the intestinal mucosa, expanding the and distorting the villi. The affinity of the T-cells to the intestinal mucosa is the basis for their epitheliotropism. However, neoplastic cells may also invade the entirety of the gastrointestinal wall causing perforation. Typical immunophenotypic profile of MEITL shows CD2+, CD3+, CD4−, CD5−, CD7+, CD8+, and CD56+. Granzyme expression is less consistent.[12] Differential diagnoses are listed below (Table 1). The closest differential diagnosis with the case presented is EATL, which shares similar cytotoxic T-cell immunophenotype. However, for EATL, tumor cells are more polymorphic, and CD8 and CD56 are negative. Intestinal T-cell lymphoma, NOS, should also be considered which can also show atypical lymphoid infiltration and epitheliotropism. However, this can be excluded with its CD4 positivity and CD56 negativity.

Another entity that should be excluded is Extranodal NK/T-cell lymphoma (ENKTL). These tumors are composed of medium-sized lymphoid cells, with positivity for CD3, Granzyme B, and variably express CD56, similar to our case. However, no angiocentric or angioinvasive pattern was seen, which is characteristic of ENKTL.[13] Epstein Barr Virus positivity for this case is still in question, however, since the patient was lost to follow up, further testing cannot be done. It is therefore recommended that EBER-In Situ Hybridization be performed to complete rule out this entity, as there may be a modification in the chemotherapy regimen. Addition of L-Asparaginase to the traditional CHOP regimen is recommended in some studies.[14] Other studies recommend a different treatment altogether, without the use of anthracycline-based drugs, using asparaginase-gemcitabine based regimens.[15]

Etiology of MEITL remains unknown. Neoplastic T-cells are implicated to arise from intestinal intraepithelial lymphocytes with CD8+ and CD56+. Whole genome

Table 1. Differential diagnoses

	Patient	Monomorphic Epitheliotropic Intestinal T-cell Lymphoma	Enteropathy-Associated T-cell Lymphoma	Intestinal T-cell Lymphoma, NOS	Extranodal NK/T-cell Lymphoma
CD3	+	+	+	+	+
CD4	-	-	-	+/-	-
CD5	-	-	-	+	-
CD8	+	+	-	+	-
CD56	+	+	-	-	+/-
CD68	+	+			
CD30	-	-		-	
CD20	-	-	-	-	-
Granzyme B	+	+	+	+	+
LCA	+	+	+	+	+
CK	-	-	-	-	-

sequencing showed characteristic activating mutations of the JAK-STAT pathway, with mutations of *STAT5B* being the most common.[16] In one small study using Next Generation Sequencing, mutations in STAT5B, SETD2, NRAS, KRAS, and JAK3 were identified.[17] Fluorescence in-situ hybridization studies have also demonstrated SETD2 deletions and MYC gene locus alterations. Expression of MYC was associated with less monomorphic morphology on histology, p53 mutations, and poorer overall survival in one study.[6]

Although diagnosis rests mostly on the morphology and immunophenotypic profile, molecular studies can help in detecting more mutations which can be future drug targets. Reporting should be intensified to characterize this type of tumor and increase the index of suspicion for its occurrence before patients deteriorate. More knowledge on MEITL can help in the search for better therapeutic regimen for these patients and alter their clinical course.

CONCLUSION

This is a case of a 64-year-old male presenting with abdominal pain and ruptured viscous at the emergency room. On histopathologic examination, dense sheets of small to medium sized mononuclear cells were seen to infiltrate and perforate the bowel wall. The tumor was immunoreactive for CD3, CD8, CD56, CD68 and Granzyme B, confirming its diagnosis as Monomorphic Epitheliotropic Intestinal T-cell Lymphoma. Although this entity is commonly seen in the Asian population, no case has been reported yet in the Philippines. Reporting should be intensified to characterize this type of rare tumor to help future researchers in their search for a cure.

ETHICAL CONSIDERATION

Multiple attempts were made to obtain informed consent from the patient. However, the patient was lost to follow-up, and no responses to these communications were received.

STATEMENT OF AUTHORSHIP

The authors certified fulfillment of ICMJE authorship criteria.

DATA AVAILABILITY STATEMENT

No datasets were generated or analyzed for this study.

AUTHOR DISCLOSURE

The authors declared no conflicts of interest.

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