Truth lies below: A case report and literature review of typical appearing polyps yet with an atypical diagnosis

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**Abstract**

**BACKGROUND**

Enteropathy associated T-cell lymphoma (EATL) is a rare form of peripheral T-cell lymphoma and makes up less than 5% of gastrointestinal lymphomas. EATL can be divided into type 1 which is associated with celiac disease, and monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL), formally type 2, which is not associated with celiac disease.

**CASE SUMMARY**

We present a 60-year-old African American female, without celiac disease, who presented with abdominal pain, diarrhea, and 30 lb. weight loss over a 3 month period. She was subsequently diagnosed with EATL throughout her entire gastrointestinal tract. She is currently undergoing chemotherapy with EOCH (Etoposide, Oncovin, Cyclophosphamide, and Hydroxydaunorubicin). EATL is most common in the Asian and Hispanic population yet the incidence in African Americans is uncertain and emphasizes the rarity of this case. A literature review was included to further emphasize similarities and differences between our case and previously reported cases of MEITL.

**CONCLUSION**

The patient was diagnosed with EATL, immunochemical testing was not conclusive for MEITL however was suggestive of the disease.

**Key words:** Enteropathy associated T-cell lymphoma; Monomorphic epitheliotropic intestinal T-cell lymphoma; Peripheral T-cell lymphoma; Gastrointestinal lymphoma; Endoscopy; Case report; Literature review
INTRODUCTION

Peripheral T-cell lymphoma (PTCL) is a small subset of aggressive non-Hodgkin lymphomas (NHL)\(^1\). One of the rarer entities of PTCL is enteropathy associated T-cell lymphoma (EATL). In general, EATL is diagnosed upon immunophenotype of small intestine biopsy and is only associated with approximately 5% of gastrointestinal (GI) lymphomas and less than 1% of NHL\(^2\). EATL is most commonly found in adult populations with a high incidence of celiac disease\(^3\). However, EATL can be divided into type 1 disease, which is associated with celiac disease, and monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL), formally type 2, which is not associated with celiac disease\(^1\). MEITL is unique upon its immunophenotypic features compared to type 1 EATL\(^4\) and has a higher incidence in Asian and Hispanic populations\(^5\). Ideal treatment of EATL consist of combination chemotherapy and hematopoietic cell transplantation (HCT)\(^6\). Here we present a unique case of EATL, in an African American patient without celiac disease.

CASE PRESENTATION

Chief complaints

A 60-yr-old African American female with a remote medical history of breast cancer, status post double mastectomy, presented with 1 wk of bilateral lower abdominal pain that was associated with diarrhea, early satiety, and a 30 lb. weight loss (over 3 mo).

Diagnostic evaluation

A computed tomography (CT) scan revealed a large circumferential mass involving the transverse colon which extended approximately 14 cm in length (Figure 1 A, B, and C). On physical exam, her vitals were stable. Her abdominal exam was soft and non-distended, with mild pain to palpation to her lower abdomen. Her laboratory work-up was notable for a mild leukocytosis (white blood count 13.4 bil/L, hemoglobin 15.7 g/dL, platelets 404 bil/L), hyponatremia and acute kidney injury (sodium 123 mmol/L and creatinine 2.52 mg/dL (baseline creatinine was normal)). Her liver function tests (including albumin and protein level), lactic acid, lipase, infectious stool studies (i.e., *Clostridium difficile* infection, ova and parasites, *Shiga* toxin producing *Escherichia coli*, *Salmonella*, *Shigella*, *Campylobacter*, or *Escherichia coli* O157) were all normal.

She underwent an esophagogastroduodenoscopy (EGD) and colonoscopy. EGD was notable for non-erosive gastropathy and normal appearing duodenum for which biopsies were obtained. Colonoscopy was notable for nodular ileal and colonic mucosa with multiple colonic polyps (Figure 2). In addition, she had a 15 cm malignant appearing stricture in the transverse colon (Figure 3). The largest colonic polyp was approximately 2.5 cm in her rectum (Figure 4). Pathology results of her stomach, duodenum, terminal ileum, colon, transverse stricture and all of her colonic polyps were notable for MEITL (Figure 5). There was no evidence of celiac disease on duodenal biopsy. There was no evidence of adenocarcinoma throughout her colon.

For the patient’s newly diagnosed high grade EATL, she underwent staging bone marrow biopsy and positron emission tomography (PET)/CT scan. Bone marrow biopsy was without overt morphologic or flow cytometry evidence of T-cell
Computed tomography scan revealed a large circumferential mass involving the transverse colon which extended approximately 14 cm in length (arrows). A: Axial; B: Coronal; C: Transverse.

lymphoma or metastatic malignancy. PET/CT scan with abnormal F-18 fluorodeoxyglucose (FDG) activity associated with the transverse colonic mass, mesenteric lymphadenopathy, focal uptake within the rectum, intense uptake throughout the bone marrow, and portions of the spleen.

The patient was started on chemotherapy with EOCH (Etoposide, Oncovin, Cyclophosphamide, and Hydroxydaunorubicin). She was not a candidate for HCT given her functional status. Unfortunately, despite her aggressive chemotherapy regimen, her disease persisted. At her 6-month follow-up, her repeat PET/CT scan with abnormal FDG activity associated with the transverse colon and rectum. Repeat colonoscopy noted a large lymphoma polyploidy lesion.

**FINAL DIAGNOSIS**

The patient was diagnosed with EATL, immunochemical testing was not conclusive for MEITL however was suggestive of the disease.

**TREATMENT**

The patient underwent chemotherapy with EOCH (Etoposide, Oncovin, Cyclophosphamide, and Hydroxydaunorubicin).

**OUTCOME AND FOLLOW-UP**

At her 6-mo follow-up, her repeat PET/CT scan with abnormal FDG activity associated with the transverse colon and rectum. Repeat colonoscopy noted a large lymphoma polyploidy lesion.

**DISCUSSION**

EATL is a rare form of PTCL that generally effects middle aged Caucasians. The majority of cases involve the small intestine (up to 90%), while the stomach and colon are less commonly involved (35% and 8% respectively)\(^{[3,4]}\). Additionally, many patients with EATL have celiac disease on histology\(^{[7]}\). Our patient is unique as she is an African American with involvement of entire GI tract, and no histological evidence of celiac disease.

Endoscopically, EATL often presents with nodularity, circumferential ulcers, and at times perforation. Identifiable masses or polyps are not generally appreciated\(^{[8]}\). In this case, the colonoscopy was notable for multiple large polyps (largest being 2.5 cm in the rectum) and a malignant appearing stricture.

Ideal treatment of EATL is consisted of combination chemotherapy and HCT. However, if patients are with poor functional status, HCT may not be an option. Sieniawski et al\(^{[6]}\) demonstrated in a retrospective study that patients with EATL undergoing chemotherapy (ifosfamide, etoposide, epirubicin, and methotrexate) followed by HCT had progression free survival and overall survival (OS) at 5 yr of
52% and 60%, respectively. Chemotherapy alone has a much poorer prognosis with 5 yr OS of 10%-20%[8].

To review previous cases, we also performed a PubMed literature search. The search was conducted using the phrase “case report” and “monomorphic epitheliotropic intestinal T-cell lymphoma”. Six articles were identified between 2016 and 2018, involving nine patients. The majority of cases reported were in Southeast Asia. The patient’s ages ranged from 40 to 83 yr old and the majority of cases were male (66%). The most common presenting symptoms were diarrhea (44%) and weight loss (33%). Interestingly, one patient presented with severe dyspnea. The patient was noted to have a pleural effusion secondary to MEITL. Treatment regimens mainly consisted of various chemotherapy regimens with 33% of patients receiving stem cell transplants. Unfortunately, survival after diagnosis was poor and ranged from 5 d to 3 yr despite therapy (Table 1)[9-14].

**CONCLUSION**

In conclusion, our patient continues to undergo therapy. Her subsequent PET scan and endoscopy have identified persistent disease. Our case is limited in that the patient has yet to complete chemotherapy. At her 6-mo follow-up, her repeat PET/CT scan with abnormal FDG activity associated with the transverse colon and rectum. Repeat colonoscopy noted a large lymphoma polyploidy lesion. There is no gold standard for monitoring EATL and repeat endoscopies can be decided on a case-by-case basis with guidance from oncology and further repeat imaging.
Table 1  Literature review of case reports of monomorphic epitheliotropic intestinal T-cell lymphoma

| Author               | Year and Location | Patient Age and Gender | Presenting symptom                  | History of celiac disease? | Treatment                                      | Prognosis after diagnosis |
|----------------------|-------------------|------------------------|-------------------------------------|---------------------------|-----------------------------------------------|---------------------------|
| Chen et al\(^9\)     | 2016, Singapore   | 60 y/o, Male           | Abdominal pain                      | No                        | CHOP + IVE/MTX + autologous stem cell transplant | Deceased at 2 wk           |
| Ishibashi et al\(^10\) | 2016, Japan       | 60 y/o, Male           | Persistent diarrhea and weight loss  | No                        | CHASE and autologous stem cell transplant      | Deceased at 3 yr          |
| Ishibashi et al\(^10\) | 2016, Japan       | 40 y/o, Female         | Diarrhea and weight loss            | Not stated                | THP-COP                                       | Deceased at 2 mo          |
| Ishibashi et al\(^10\) | 2016, Japan       | 50 y/o, Female         | Abdominal distention                | Not stated                | CHOP + high dose MTX/cytarabine + allogeneic stem cell transplant | Deceased at 9 mo          |
| Ishibashi et al\(^10\) | 2016, Japan       | 70 y/o, Male           | Nausea                              | Not stated                | SMILE                                         | Deceased at 9 mo          |
| Aiempapanakit et al\(^11\) | 2017, Thailand   | 67 y/o, Male           | Chronic diarrhea and weight loss     | Not stated                | Anthracyline based chemotherapy               | Deceased at 2 mo          |
| Antoniadou et al\(^12\) | 2016, Greece     | 76 y/o, Male           | Severe dyspnea                      | Not stated                | Unable to tolerate treatment and deceased     | Deceased at Day 5         |
| Aoyama et al\(^13\)  | 2018, Japan       | 83 y/o, Male           | Fever and diarrhea                  | Not stated                | CHOP followed by DeVIC                        | Deceased yet no timeframe stated |
| Pan et al\(^14\)     | 2018, Taiwan      | 76 y/o, Female         | Intermittent abdominal pain         | No                        | CEOP                                          | Deceased at 3.7 mo        |

y/o = years-old; CHOP = cyclophosphamide, doxorubicin, vincristine, and prednisone; IVE = ifosfamide, vincristine, and etoposide; MTX = methotrexate; CHASE = cyclophosphamide, cytarabine, etoposide, and dexamethasone; THP-COP = pirarubicin, cyclophosphamide, vincristine, and prednisolone; SMILE = dexamethasone, methotrexate, ifosfamide, L-asparaginase, and etoposide; DeVIC = etoposide, doxorubicin, oncovin, and prednisolone; CEOP = cyclophosphamide, epirubicin, vincristine, and prednisolone.

Figure 3 Colonoscopy with 15 cm malignant appearing stricture in the transverse colon.

Figure 4 Colonoscopy. 2.5 cm in her rectum polyp.
colon, transverse stricture and all of her colonic polyps are consistent with enteropathy associated T-cell lymphoma. Prominent intraepithelial lymphocytosis is present (arrows; Hematoxylin and Eosin, 500 ×). Pathology results of her stomach, duodenum, terminal ileum, colon, transverse stricture and all of her colonic polyps are consistent with enteropathy associated T-cell lymphoma.

Figure 5  The infiltrate is composed of small to intermediate-sized lymphocytes with round nuclei, inconspicuous nucleoli and lacks an inflammatory background. Prominent intraepithelial lymphocytosis is present (arrows; Hematoxylin and Eosin, 500 ×). Pathology results of her stomach, duodenum, terminal ileum, colon, transverse stricture and all of her colonic polyps are consistent with enteropathy associated T-cell lymphoma.

REFERENCES

1  Swerdlow SH, Campo E, Pileri SA, Harris NL, Stein H, Siebert R, Advani R, Ghielmini M, Salles GA, Zelenetz AD, Jaffe ES. The 2016 revision of the World Health Organization classification of lymphoid neoplasms. Blood 2016; 127: 2375-2390 [PMID: 26980727 DOI: 10.1182/blood-2016-01-643569]

2  Zettl A, deLeeuw R, Haralambevia E, Mueller-Hermelink HK. Enteropathy-type T-cell lymphoma. Am J Clin Pathol 2007; 127: 701-706 [PMID: 17511112 DOI: 10.1309/NW2BK1DX8IEOQ551]

3  Cellier C, Delabesse E, Helmer C, Patey N, Matsuchansky C, Jabri B, Macintyre E, Cerf-Bensussan N, Brousse N. Refractory sprue, coeliac disease, and enteropathy-associated T-cell lymphoma. French Coeliac Disease Study Group. Lancet 2000; 356: 203-208 [PMID: 10963198 DOI: 10.1016/S0140-6736(00)02481-8]

4  Delabie J, Holte H, Vose JM, Ullrich F, Jaffe ES, Savage KJ, Connors JM, Rimsza L, Harris NL, Müller-Hermelink K, Rüdiger T, Cotifler B, Gascoyne RD, Berger F, Tobinai K, Au WY, Liang R, Meibesrat E, Hochberg EP, Pileri S, Fedetcio M, Nathwani R, Armitage JO, Weisenburger DD. Enteropathy-associated T-cell lymphoma: clinical and histological findings from the international peripheral T-cell lymphoma project. Blood 2011; 118: 148-155 [PMID: 21566094 DOI: 10.1182/blood-2011-02-355210]

5  Deleeuw RJ, Zettl A, Klinker E, Haralambevia E, Trottier M, Char I, Ge Y, Gascoyne RD, Chott A, Müller-Hermelink HK, Lam WL. Whole-genome analysis and HLA genotyping of enteropathy-type T-cell lymphoma reveals 2 distinct lymphoma subtypes. Gastroenterology 2007; 133: 1902-1911 [PMID: 17484883 DOI: 10.1053/j.gastro.2007.03.036]

6  Sieniawski M, Angamuthu N, Boyd K, Chastty R, Davies J, Foresyth P, Jack F, Lyons S, Mounter P, Revell P, Proctor SJ, Leonard K. Evaluation of enteropathy-associated T-cell lymphoma comparing standard therapies with a novel regimen including autologous stem cell transplantation. Blood 2010; 115: 3664-3670 [PMID: 20197551 DOI: 10.1182/blood-2009-07-231324]

7  Isaacson PG, O’Connor NT, Spencer J, Bevan DH, Connolly CE, Kirkham N, Pollock DJ, Waincoat JS, Stein H, Mason DY. Malignant histiocytosis of the intestine: a T-cell lymphoma. Lancet 1985; 2: 688-691 [PMID: 2865677 DOI: 10.1016/s0140-6736(85)90081-2]

8  Gale J, Simmons PD, Mead GM, Sweetenham JW, Wright DH. Enteropathy-type intestinal T-cell lymphoma: clinical features and treatment of 31 patients in a single center. J Clin Oncol 2000; 18: 795-803 [PMID: 10673532 DOI: 10.1200/JCO.2000.18.4.795]

9  Chen Y, Tan SY, Petersson BF, Khor YM, Gopalakrishnan SK, Tan D. Occult recurrence of monomorphic epitheliotropic intestinal T-cell lymphoma and the role of MATK gene expression in diagnosis. Hematol Oncol 2017; 35: 852-855 [PMID: 26948059 DOI: 10.1016/j.hemonc.2017.01.001]

10  Ishibashi H, Nimura S, Kayashima Y, Takamatsu Y, Aoyagi K, Harada N, Kadowaki M, Kamio T, Sakisaka S, Takeshita M. Multiple lesions of gastrointestinal tract invasion by monomorphic epitheliotropic intestinal T-cell lymphoma, accompanied by duodenal and intestinal enteropathy-like lesions and microscopic lymphocytic proctocolitis: a case series. Diagn Pathol 2016; 11: 66 [PMID: 27457239 DOI: 10.1186/s13000-016-0519-x]

11  Aiempbanaki K, Amatawet C, Chiratikarnwong K, Auepemkiate S, Kayasut K, Suwiwat S, Apinantriyo B. Erythema multiforme-like cutaneous lesions in monomorphic epitheliotropic intestinal T-cell lymphoma: a rare case report. J Cutan Pathol 2017; 44: 183-188 [PMID: 27862162 DOI: 10.1111/cup.12864]

12  Antoniadou F, Dimitrakopoulou A, Voutsinas PM, Vrettou K, Vlahadami I, Voulgarelis M, Korkolopoulou P, Kafasi N, Mikou P. Monomorphic epitheliotropic intestinal T-cell lymphoma in pleural effusion: A case report. Diagn Cytopathol 2017; 45: 1050-1054 [PMID: 28681573 DOI: 10.1002/dc.23772]

13  Aoyama Y, Tsunemine H, Zushi Y, Maruoka H, Goto Y, Kodaka T, Itoh T, Takahashi T. Colonial monomorphic epitheliotropic intestinal T-cell lymphoma with novel phenotype of cytoplasmic CD3 expression. J Clin Exp Hematop 2018; 58: 102-106 [PMID: 29657256 DOI: 10.3960/j.jeh.18002]

14  Pan ST, Ko YH, Tan SY, Chuang SS. Primary cutaneous peripheral T-cell lymphoma with a late relapse solely in the ileum mimicking monomorphic epitheliotropic intestinal T-cell lymphoma. Pathol Res Pract 2018; 214: 2106-2109 [PMID: 30477646 DOI: 10.1016/j.prp.2018.10.002]

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