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Case report

Intestinal presentation of non-Hodgkin lymphoma: Case report☆

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ABSTRACT

Background: MEITL (monomorphic epitheliotropic intestinal T-cell lymphoma) is a rare primary intestinal T-cell lymphoma associated with high mortality rate. Being intestinal this implies difficult diagnostic workup and management.
Case: We present a case of 59-year-old male presented with abdominal pain and found to have 15 × 11 cm mass in the right iliac fossa. Decision for surgical excision was retained and the Pathological examination determined monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) with CD30 positive immune-histochemical profile.
Conclusion: MEITL is a rare entity which retains challenging diagnosis and management as well as variable immune-histochemical profile. In the absence of clear guidelines for the management of intestinal manifestation of lymphoma, surgical approach may have its indications.

1. Introduction

Peripheral T-cell lymphomas (PTCL) are rare and fast-growing types of non-Hodgkin lymphoma associated with high fatality rate. Precisely, PTCL involves the intestine and this enteropathy subtype has a 4.7% incidence rate and a 5 years overall survival of 20% [1,2]. These neoplasms consist of 29 distinct entities [1]. Prior to the new classification of WHO this type was identified as one entity but divided into 2 subgroups: Enteropathy associated T-cell lymphoma (EATL) and monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) [3,4]. Moreover, MEITL is an uncommon primary intestinal T-cell lymphoma associated with high mortality rate and male predominance [5]. It affects the small bowel, mainly the jejunum and less likely the large bowel and stomach [5]. The clinical picture can be unspecific ranging from subtle onset characterized by weight loss, abdominal discomfort, changes in bowel habits, diarrhea or rectal bleeding to acute presentation with bowel perforation or obstruction [6]. In the setting of these clinical manifestations, diagnosis is challenging although early diagnosis is of high priority. We describe a unique case of MEITL presenting as a large abdominal mass. This case was reported in accordance with the SCARE 2020 criteria [7]. Written and informed consent was taken from the patient to publish this paper and its accompanying images.

2. Case

We present a case of 59-year-old male patient known to have hypertension, presented for a 3-month history of 10 kg weight loss without intention, recurrent nocturnal episodes of fever, night sweats, and abdominal pain. The patient has no relevant family history and he was not on specific medications other than anti-hypertensive ones. Upon admission, physical exam was remarkable for abdominal mass on palpation in the right iliac fossa. Moreover, routine blood workup showed a microcytic anemia with a hemoglobin of 9.5 g/dl. Abdominal CT scan showed a large lobulated, relatively well circumscribed heterogeneous soft tissue mass in the lower abdomen and right iliac fossa measuring 15 × 11 cm diameter engulfing the ileum, appendix, bladder and adherent to the sigmoid as shown by (Fig. 1). Notably, the patient was opted for gastroscopy and colonoscopy which were negative.

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We opted for surgery by laparotomy by midline incision. Perioperative stenting of the ureters was secured. The mass was adherent to the small bowel, appendix, urinary bladder and the sigmoid. Excision of the mass en-bloc was achieved by resection and anastomosis of 20 cm of the ileum and partial cystectomy. In the postoperative period the patient benefited from one day of surveillance in the intensive care unit and then was transferred to the regular floor. The antibiotic coverage was extended till 48 h post op. Recuperation of the bowel function and the food intake was progressive and without difficulties. Moreover, the abdominal drain was removed at day 3 post op. Respiratory-wise, the patient used the spirometry regularly and he was always afebrile. On biological level, the lab tests of control were within normal limits. Following this favorable post operative state, the patient was authorized leaving the hospital on day 7 post op with uneventful stay. Furthermore, pathology report revealed the presence of poorly differentiated cells. These cells were isolated medium to large- sized with circonvuluted hyperchromatic nuclei along with zone of coagulation necrosis and tumoral necrosis. Regarding immuno-histo-chemistry, the tumor was CD30+, CD3+, ALK1-, CD43-, EMA-as shown in (Fig. 2). There were no lymph nodes involved histologically. Furthermore he underwent bone marrow biopsy showing no bone marrow involvement, thus the final Ann Arbor stage was IE.

Hence the diagnosis was in favor of monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL). Besides, tissue transglutaminase IgA test was negative, ruling out celiac disease. Workup of extension was completed by PET scan showing no evidence of metastasis.

Afterwards, he completed six courses of adjuvant chemotherapy according to the following regimen: Brentuximab vedotin in combination with cyclophosphamide, doxorubicin, and prednisone. This was followed by BEAM regimen and a successful autologous stem cell transplantation. One year followup post BMT failed to show an evidence of recurrence.

3. Discussion

MEITL is rare a disease; few cases have been reported in the literature. Thus, fixed criteria are not yet established making the diagnosis challenging. This case we are reporting is atypical in that it disclaims many known features of MEITL, hence expanding the diagnostic spectrum. Concerning the immuno-histochemical profile, MEITL is known to be CD30- [8], contrary to our case, reflecting an immuno-phenotypical variation. Moreover, coagulative necrosis, a hallmark finding in EATL, is unusual in MEITL [5], however it is present in our case. On an endoscopic basis, many cases demonstrated multiple ulcerations in the small bowel associated with velvety mucosa that may cause obstruction [9] which contracts with our case where the ileo-colonoscopy was normal, thus questioning the role of endoscopy in early diagnosis. On the other hand, there are no specific guidelines for the treatment of MEITL. Current management consists of combination of surgical excision, followed by advujant chemotherapy and an autologous stem cell transplantation [10]. Hence in the lack of clear guidelines for management, we opted for surgical excision and adjuvant chemotherapy. Early detection of localized early primary lymphoma remains the most crucial yet the most difficult aim [11]. In our case, the patient presented with non-specific clinical picture yet total excision revealed early stage of MEITL.

Brentuximab vedotin (BV) is a targeted antibody-drug conjugate (ADC) used to treat CD30 positive neoplastic cells [12]. ECHELON-2 trial showed that BV added to the combination of cyclophosphamide, doxorubicin, and prednisone (BV + CHP) reduced the risk of disease progression by 29% and mortality by 34% for patients with PTCLs, compared with treatment involving a standard regimen of CHOP [13]. Knowing the aggressiveness of this type of lymphoma, high dose chemotherapy (HDT) according to the BEAM regimen was administered to the patient. This was followed by autologous stem cell transplant (ASCT) as HDT/ASCT can improve progression-free survival specially if applied during first remission [14].

4. Study limitations

This is a case report that questions the surgical indication in managing MEITL in the absence of clear guidelines for the management of this entity. However more research is needed to endorse our approach. Furthermore, this paper needs further immunohistochemical-marker analysis.

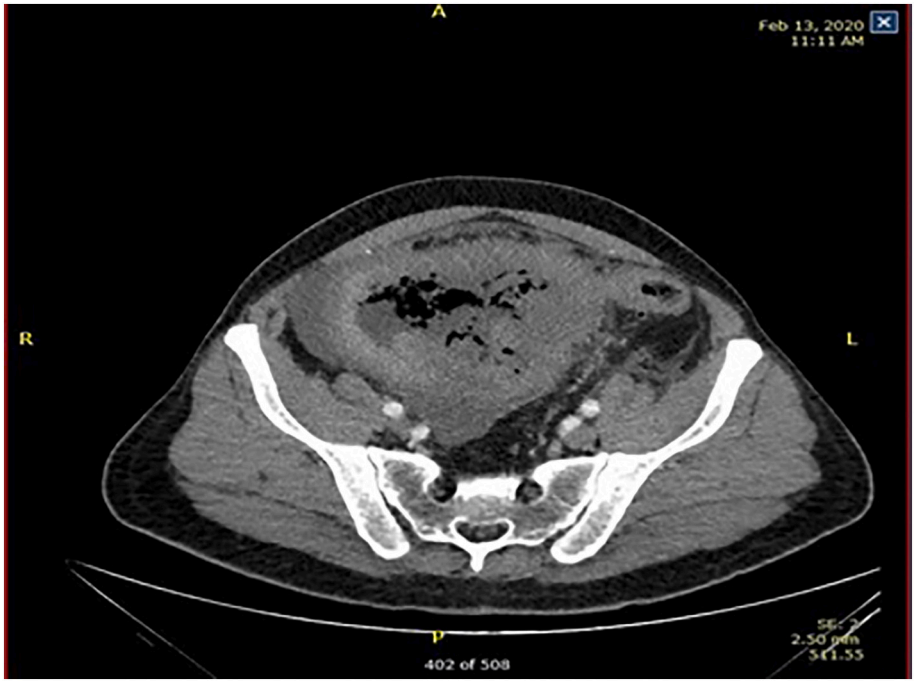

Fig. 1. Showing the abdominal mass in contact with the small bowel bladder and the sigmoid.

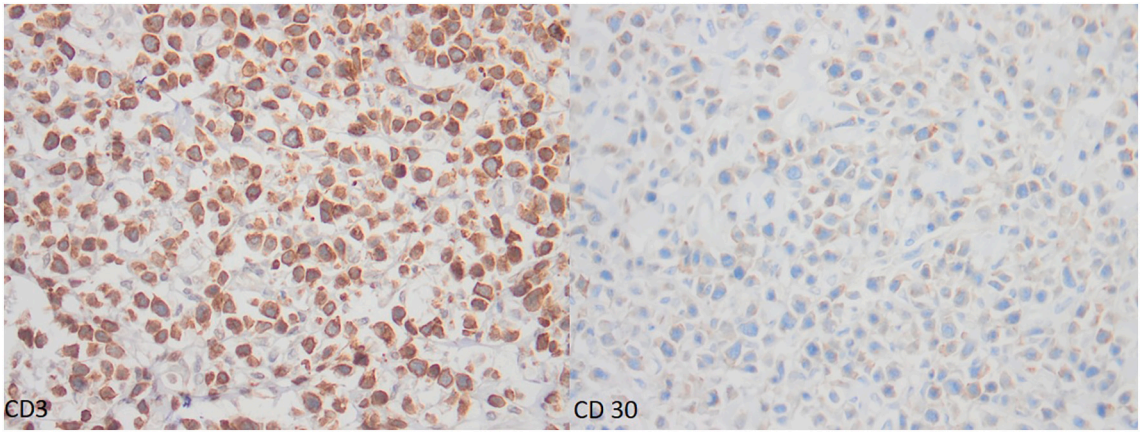

Fig. 2. Microscopic view showing CD3 and CD30 cells (×40 magnification).

5. Conclusion

Intestinal manifestation of lymphoma is a rare entity. This report highlights the surgical indication in establishing the diagnosis and the management. Moreover, MEITL may present with variable immune-histochemical profile being CD30 positive.

Informed consent

The patient provided us written informed consent to publish this case report and its accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

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CRediT authorship contribution statement

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Declaration of competing interest

None.

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Abbreviations

PTCL: Peripheral T-cell lymphomas
EATL: Enteropathy associated T-cell lymphoma
MEITL: Monomorphic epitheliotropic intestinal T-cell lymphoma
CHP: Cyclophosphamide, doxorubicin, and prednisone
CHOP: Cyclophosphamide, doxorubicin, vincristine, and prednisone
BEAM: Carmustine, etoposide, aracytin and melphalan
BMT: Bone marrow transplant
BV: Brentuximab vedotin
HDT: High dose chemotherapy
ASCT: Autologous stem cell transplant
CD: Cluster of differentiation
ALK: Anaplastic lymphoma kinase
EMA: Epithelial membrane antigen