Elmer Press Case Report J Hematol. 2014;3(3):89-90

Lennert's Maze Game: A Diagnostic Hematological Challenge

Nilma Malik[a, b], Vishwanath Pattan[a], Waqas Jehangir[a], Hongxiu Luo[a], Yazan Vwich[a], Teena Mathew[a], Shuvendu Sen[a], Abdalla Yousif[a]

Abstract

Lennert lymphoma is a subset of peripheral T-cell lymphoma (PTCL) that constitutes about 30% of T-cell lymphomas. The immunophenotypes usually are positive for pan-antigens, such as CD2, CD3, CD5 or CD7. About half cases present aberrantly without the pan-antigens. This rare case opens up a new horizon of thought process, as it illustrates the clinical presentation of PTCL with aberrant hematological parameters. It emphasizes on diagnostic and treatment challenges of Lennert lymphoma presenting with a triad of eosinophilia, lactic acidosis and multi-organ thrombosis. Early treatment with chemotherapy rather than supportive measures is crucial as it has shown significant morbidity benefits.

Keywords: Lennert; Lactic acidosis; Eosinophilia; Thrombosis; Hemophagocytosis; Chemotherapy; Thrombomodulin; Prognosis

Introduction

The incidence of peripheral T-cell lymphomas (PTCL), not otherwise specified (NOS) has increased in USA from 0.1 cases/100,000 population in 1992 to 0.4 cases/100,000 population in 2006 [1]. We report a rare case of Lennert's, variant of PTCL presenting with eosinophilia, multi-organ thrombosis and lactic acidosis.

Case Report

A 47-year-old male was admitted for evaluation of 4-day history of worsening shortness of breath and pain in his right hand and leg. One month prior to admission he was diagnosed with bilateral lower extremity, hepatic vein and inferior vena cava thrombosis and was started on warfarin. There was no family history of venous clots or stroke. Upon admission he had an HR 87/min, BP 125/67 mm Hg, RR 16/min, T 98.6 °F and SO_2 95%. Clinical examination revealed absent right ulnar and dorsalis pedis pulses and a papular rash over trunk and arms (Fig. 1a, b). Labs were significant for Hb 7 g/dL, WBC 19,800/µL (neutrophil 82%, eosinophils 69% and metamyelocyte 1%). Platelets were 487,000/µL, glucose 120 mg/dL, BUN 39 mg/dL, ceatinine 0.8 mg/dL, bicarbonate 14 mEq/L, anion gap 22 mEq/L, lactic acid 6.1 mmol/L and INR 1.8. Urine drug screen, sepsis and hypercoagulable workup were negative. Peripheral blood smear showed anisopoikilocytosis, and no parasites were identified. CT of chest, abdomen and pelvis showed no organomegaly, lymphadenopathy or hemorrhage. Right lower and upper extremity angiography revealed ulnar and popliteal artery thrombosis. The patient was transfused blood and treated with t-PA. Nevertheless he underwent amputation of his lower limb and right index and ring fingers. Dilemma remained as despite of adequate treatment patient leukocytosis, eosinophilia and lactic acidosis, persisted.

Since there was a high likelihood of hematological malignancy the patient underwent extensive workup. Bone marrow PCR gene rearrangement study revealed the presence of clonal population of T cells, consistent with T-cell leukemia. Immunophenotype staining showed CD 45+, 38+, 34+ and 117+. Skin biopsy revealed clusters of epithelioid cells intermingled with small lymphocytes (Fig. 1c). He was started on cyclophosphamide, doxirubucin, vincristine and prednisone. Chemotherapy resulted in resolution of rash and lactic acidosis and improvement of the hematological picture.

Discussion

Lennert lymphoma is a subtype of PTCL, NOS, which accounts for 6% of non-Hodgkin lymphomas. The immunophenotype of PTCL varies greatly and demonstrates expression of pan-T-antigens (CD2, CD3, CD5 or CD7). Roughly half cases present aberrantly with loss of pan-antigens [2],

Manuscript accepted for publication July 25, 2014

[a]Internal Medicine Department, Raritan Bay Medical Center, 530 New Brunswick Avenue, Perth Amboy, NJ 08861, USA
[b]Corresponding Author: Nilma Malik, 97 Tennyson Street, Carteret, NJ 07008, USA. Email: nilmamalik1@gmail.com

doi: http://dx.doi.org/10.14740/jh165w

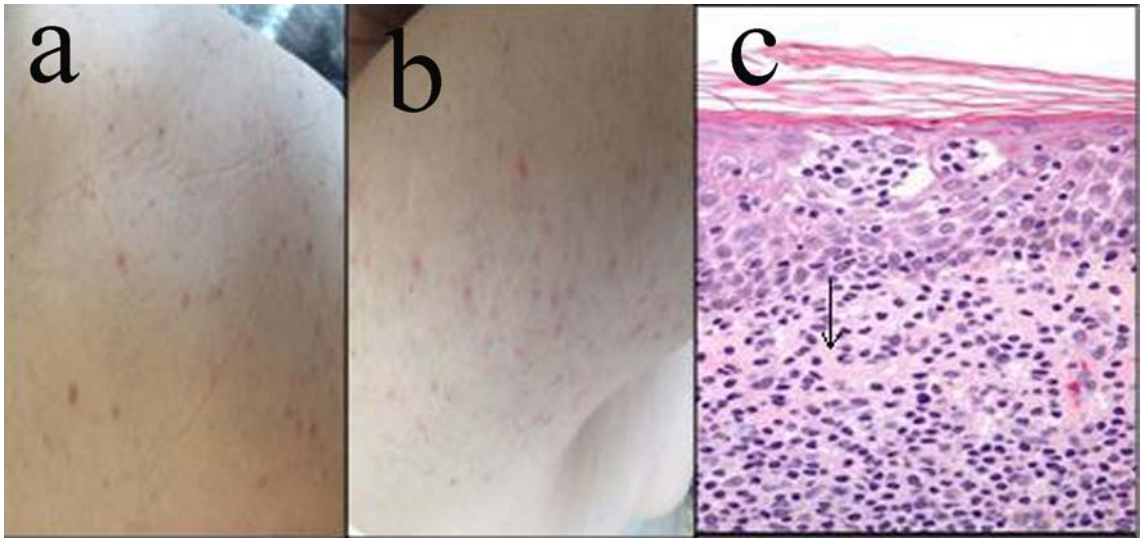

Figure 1. (a, b) Rash associated with Lennert lymphoma. (c) Histology of Lennert lymphoma, arrow showing epithelioid cell cluster.

as in our patient. Clinical presentation includes lymphadenopathy, systemic B symptoms, eosinophilia or hemophagocytosis.

Eosinophilia occurs as a reaction to leukemia and has been reported as rare cause of arterial and venous thrombosis. The thrombotic event is induced by an eosinophile, which inhibits thrombomodulin function. Thrombomodulin serves as a cofactor for activation of protein C. Thus deficiency of activated protein C engenders a prothrombotic environment. This can explain the multi-organ thrombosis in our patient despite of having a therapeutic INR, as assessment of PT does not reflect levels of endogenous anticoagulants such as protein C [3].

Type B lactic acidosis is related to increased tumor burden and is associated with acute exacerbation of leukemias. Lactic acidosis originating from malignancies does not respond to usual resuscitative measures such as fluids and antibiotics and resolves only with chemotherapy.

Conclusion

The current poor prognostic factors for PTCL include age > 60, elevated serum lactate dehydrogenase, Eastern Cooperative Oncology Group (ECOG) Performance Status > 2 and bone marrow involvement [4]. Considering the wide spectrum of presenting symptoms, progressive nature and frequent relapses of the disease, it is thus imperative to have high index of clinical suspicion for "Lennert lymphoma" as early treatment has significant morbidity and mortality benefits. Also it may be a reasonable thought to widen the set of hematological prognostic factors for PTCL, NOS such as "lactic acidosis, eosinophila and leukocytosis" as in our patient.

Funding

None.

Conflict of Interest

None.

References

1. Abouyabis AN, Shenoy PJ, Lechowicz MJ, Flowers CR. Incidence and outcomes of the peripheral T-cell lymphoma subtypes in the United States. Leuk Lymphoma. 2008;49(11):2099-2107.
2. Weiss LM, Crabtree GS, Rouse RV, Warnke RA. Morphologic and immunologic characterization of 50 peripheral T-cell lymphomas. Am J Pathol. 1985;118(2):316-324.
3. Tripodi A, Mannucci PM. The coagulopathy of chronic liver disease. N Engl J Med. 2011;365(2):147-156.
4. Gallamini A, Stelitano C, Calvi R, Bellei M, Mattei D, Vitolo U, Morabito F, et al. Peripheral T-cell lymphoma unspecified (PTCL-U): a new prognostic model from a retrospective multicentric clinical study. Blood. 2004;103(7):2474-2479.

