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CASE REPORT

Primary Mediastinal B-cell Lymphoma Presenting as Chest Pain in a Young Woman and Treated with EPOCH-R☆

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Abstract

Primary mediastinal B-cell lymphoma (PMBCL) is a rare type of diffuse large B-cell lymphoma, constituting 2−4% of non-Hodgkin lymphomas. Here we present a case of PMBCL initially presenting as pleuritic chest pain in an otherwise healthy 33-year-old woman. PMBCL typically presents as a large, fast-growing tumor limited to the mediastinum, making the iliosacral involvement in this case unusual. R−CHOP is the most commonly used chemotherapy regimen, although more aggressive therapies like EPOCH-R can spare the need for consolidative radiation therapy. PMBCL represents one of the greatest diagnostic and therapeutic challenges in malignant hematology.

Keywords: Non-hodgkin lymphoma, Diffuse large B-Cell lymphoma, R−CHOP, EPOCH-R, Chest pain

1. Introduction

Primary mediastinal B-cell lymphoma (PMBCL) is a type of diffuse large B-cell lymphoma thought to be derived from the thymic B cell.[1] It presents a diagnostic challenge due to its overlapping characteristics with other lymphomas.[2] Here we present a case of primary mediastinal B-cell lymphoma in a young woman treated with EPOCH-R.

2. Case

A 33-year-old female with no significant past medical history presented to the emergency department with sharp, left-sided pleuritic chest pain of 3 weeks' duration. Initial work-up revealed elevated D-dimer. CTA of the chest was negative for pulmonary embolism but did show heterogeneous soft tissue within the anterior and superior mediastinum with associated lymphadenopathy. AFP and beta-HCG were normal. LDH was elevated at 256 U/L. At that time it was felt the mass would be difficult to biopsy by either mediastinoscopy or bronchoscopy, and she was discharged with a plan for repeat scanning in 4 weeks.

Two weeks later she re-presented with worsening pleuritic chest pain though now associated with shortness of breath and rib pain. A repeat CTA of the chest showed enlarged mediastinal lymphadenopathy and lytic lesions involving multiple ribs with adjacent soft tissue thickening suspicious for neoplastic involvement.

Nuclear bone scan showed abnormal uptake within the ribs containing the lytic lesions (Fig. 1). Left seventh rib core biopsy revealed findings most consistent with diffuse large B-cell lymphoma (Figs. 2 and 3). CD30 was weakly positive. A fluorescence in situ hybridization panel was negative for BCL-2, BCL-6, or MYC rearrangement. Bone marrow biopsy was negative for malignancy. PET scan revealed hypermetabolic activity in the anterior mediastinum, ribs, sacrum, and iliac crest.

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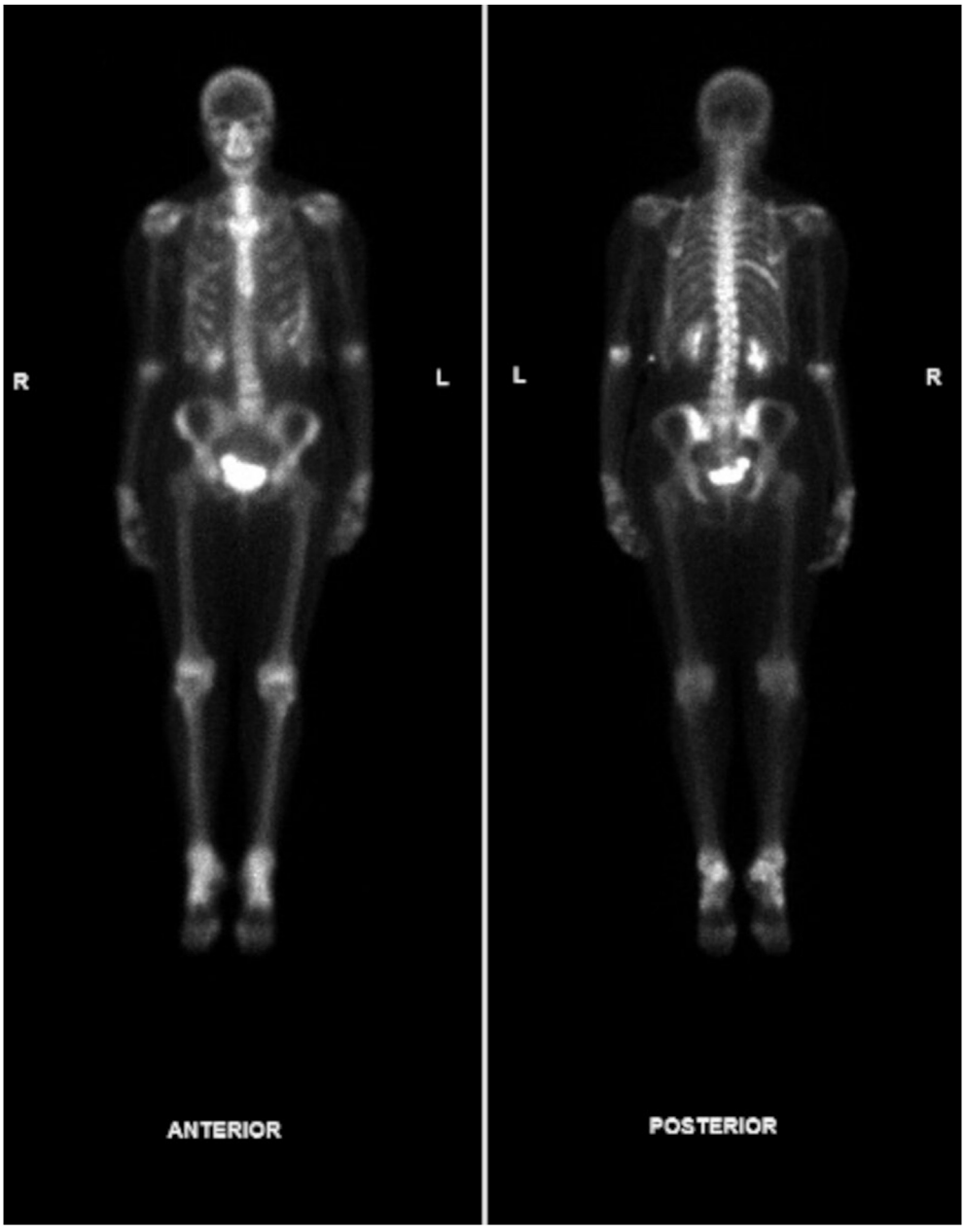

Fig. 1. Nuclear bone scan showing abnormal uptake in the posterior right ninth and anterior left seventh ribs.

A final diagnosis of primary mediastinal B-cell lymphoma, stage IV, International Prognostic Index (IPI) 3/5 was made.[3] Following hospital discharge she was seen by hematology and treatment with EPOCH-R was initiated. All symptoms of shortness of breath and rib pain were entirely resolved before completion of the first cycle.

3. Discussion

PMBCL is a rare lymphoma subtype and constitutes approximately 2–4% of all non-Hodgkin lymphomas.[4] The current WHO classification distinguishes this lymphoma as a separate entity due to its specific clinical and pathological features.[5] This disease affects mainly young adults (median age 35), predominantly women.[6] PMBCL typically presents as a large, fast-growing tumor with invasion usually limited to the anterior superior mediastinum, making the iliac and sacral involvement in our case unusual and indicative of particularly rapid spread.[7] Common signs/symptoms are cough, chest pain, dysphagia, and tachypnea, which are related to tumor mass infiltration or compression.

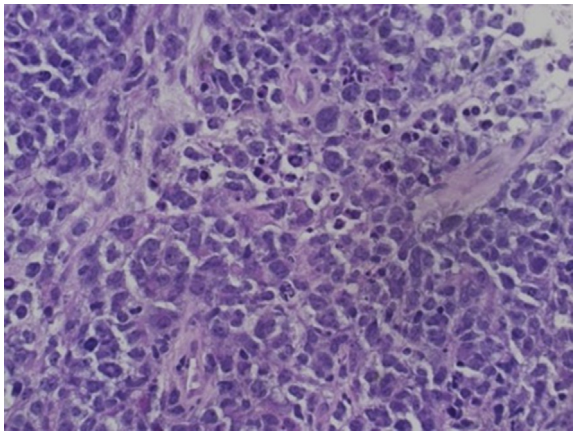

Fig. 2. Left seventh core rib biopsy showing sheet-like proliferation of large, atypical cells with frequent apoptosis, 400X.

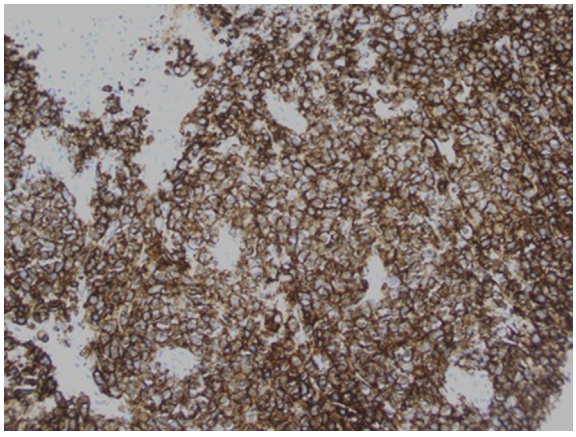

Fig. 3. CD20 immunohistochemical stain showing B lymphocytes, 200X.

Diagnosis is made by clinical presentation in conjunction with histopathological examination and immunohistochemical staining. Initial treatment is critical in the management of PMBCL since outcomes in relapsed or progressive disease are notably worse.[8–10] R–CHOP is the most commonly used chemotherapy regimen, although there are some reports on greater efficacy with more aggressive regimens.[4,10,11] More intense therapies such as EPOCH-R have been shown to spare the need for consolidative radiation therapy, hence why it was chosen in the case of this patient.[10,12–15]

4. Conclusion

In summary, PMBCL is a rare and aggressive B-cell lymphoma that affects young people and represents one of the greatest diagnostic and therapeutic challenges in malignant hematology.

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Conflict of interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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