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A RARE CASE OF LARGE B-CELL LYMPHOMA PRESENTING AS A LARGE PERICARDIAL EFFUSION WITH TAMPONADE PHYSIOLOGY

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INTRODUCTION

We highlight an interesting case of a middle-aged gentleman presenting with a rare cause of a pericardial effusion, complicated by impending cardiac tamponade. Our workup revealed a diagnosis of fluid overload-associated large B-cell lymphoma (FOLBCL), a newly defined WHO lymphoma entity. We will discuss the defining characteristics for this lymphoma, the current associated studies, and treatment.

CASE PRESENTATION

A previously healthy 60-year-old man presented with subacute exertional dyspnea, low grade fevers and chills, non-productive cough, anasarca, and a 21-lb weight gain. On admission, he was afebrile, tachycardic, and normotensive with a narrow pulse pressure. Presenting ECG noted low voltage and mild variation in QRS amplitude (**Figure 1**). Bedside point-of-care ultrasound identified a large pericardial effusion with features of tamponade, with confirmation on formal TTE (**Figure 2**). Pericardiocentesis was performed with removal of 1L dark sanguinous pericardial fluid.

Fluid yielded 4178 nucleated cells (48% lymphocytes) with many atypical lymphocytes on pathology review. Flow cytometry was positive for a lambda light chain restricted B-cell population that expressed CD45, CD19, CD20, suggesting involvement by large B-cell lymphoma. The cells were negative for CD3, CD10, CD30, HHV8 and EBV in-situ hybridization. Peripheral venous flow cytometry was negative for a monomorphic B-cell population.

PET CT noted mild increased activity involving pericardium only (**Figure 3**), no masses or lymphadenopathy. Bone marrow biopsy revealed hypercellular bone marrow with no evidence of large B-cell lymphoma.

He started chemotherapy with DA-EPOCH-R (Dose Adjusted - Etoposide + Prednisone + Vincristine + Cyclophosphamide + Doxorubicin - Rituximab). He was discharged home to continue outpatient treatment.

FIGURES & IMAGES

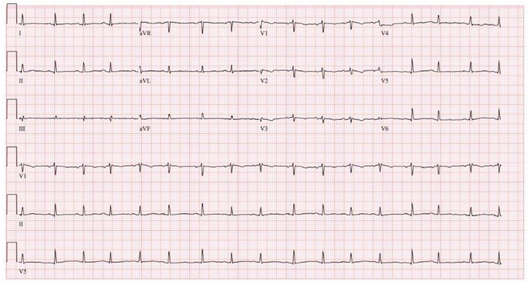


Figure 1. Presenting ECG: Sinus rhythm with sinus arrhythmia, low voltage QRS with electrical alternans

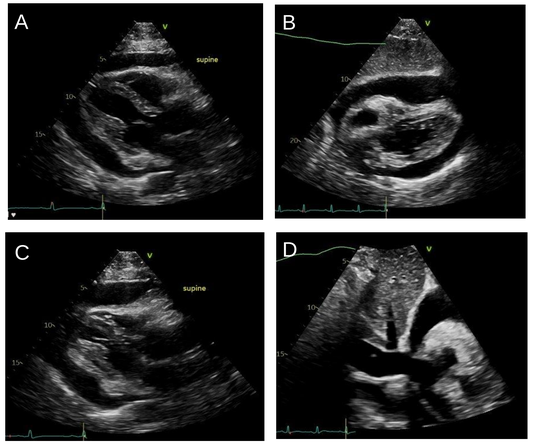


Figure 2. TTE images of presenting pericardial effusion. A: Demonstrates circumferential pericardial effusion. B/C: Demonstrates diastolic collapse of right ventricle. D: Demonstrates plethoric IVC.

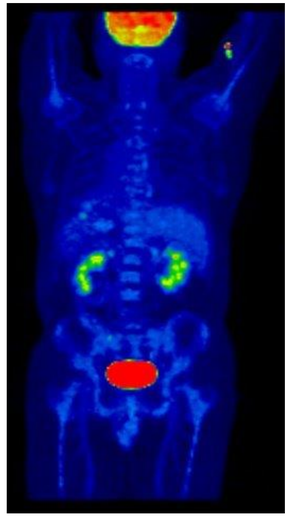


Figure 3. PET CT scan with faint activity involving the pericardium and absence of diffuse FDG uptake

DISCUSSION

- FOLBCL arises within fluid-filled cavities, without a solid component.
- Pleural, pericardial, and peritoneal cavities are involved in 75%, 34%, and 27% of cases, respectively.
- FOLBCL is most similar to primary effusion lymphoma (PEL). However, PEL is viral-associated (HHV8, EBV, +/-HIV) and exhibits pan-negative B-cell markers, whereas FOLBCL markers are positive.
- PET CT and bone marrow analysis can differentiate diffuse large B-cell lymphoma (DLBCL) from FOLBCL. DLBCL has diffuse PET-avid lymph nodes and bone marrow with lymphoma cells.
- Case reports describe treatment with CHOP (Cyclophosphamide, Doxorubicin, Vincristine, Prednisone) +/- Rituximab.
- A retrospective review of HHV8-negative PELs showed a 2-year overall survival rate of about 84%. This is a favorable prognosis when compared to the grim median overall survival of 4.8 months seen in studies for PEL.

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