Restrictive physiology masked by cardiac tamponade: A rare cooccurrence in a child with lymphoblastic T-cell lymphoma

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ABSTRACT

Cardiac involvement in T-cell lymphoma is not uncommon. Pericardial effusion is the most common manifestation of cardiac involvement with restrictive cardiomyopathy (RCM) due to tumor infiltration being extremely rare. The presence of paroxysmal nocturnal dyspnea and orthopnea in a patient presenting with pericardial effusion could be related to tamponade or underlying myocardial disease. Hence, reevaluation after pericardiocentesis is warranted. We describe a 14-year-old boy with advanced T-cell lymphoma presenting with cardiac tamponade. Repeat echocardiography after pericardiocentesis revealed mediastinal mass infiltrating cardiac chambers and great vessels along with features of RCM secondary to tumor infiltration.

Keywords: Cardiac lymphoma, restrictive cardiomyopathy, T-cell lymphoma, tamponade

INTRODUCTION

Cardiac involvement secondary to lymphoma has been reported in up to 20% of cases,[1] the majority of which are of B-cell origin.[2] T-cell lymphoma involves cardiac structures rarely and tends to be more aggressive.[2] The most common cardiac involvement detectable is pericardial effusion with rare reports of endomyocardial involvement.[3] Clinical presentation with restrictive cardiomyopathy (RCM) due to tumor infiltration is extremely rare in lymphoma.[4-6]

CLINICAL DETAILS

A 14-year-old boy presented to us with fever, weight loss, and progressive dyspnea for 2 months. At a peripheral clinic, he was found to have pleural effusion. After 1 month, he was referred to our hospital due to the lack of response, worsening dyspnea associated with new-onset orthopnea, and paroxysmal nocturnal dyspnea (PND). On arrival, he had tachypnea, tachycardia, hypotension, and pulsus paradoxus (16 mmHg). Jugular venous pressure was elevated with prominent x descent and absent y descent, the Kussmaul sign was absent, and the first and second heart sounds were muffled. There were few basal crepitations. These clinical features predominantly suggested cardiac tamponade.

Relevant blood investigations revealed elevated total leukocyte count (12,000/mm^3) and N-terminal pro-brain natriuretic peptide (NT pro-BNP) level (1255 pg/ml). Chest X-ray revealed an enlarged cardiac silhouette with evidence of pulmonary venous hypertension and bilateral mild pleural effusion. Electrocardiography revealed sinus tachycardia with T inversions in leads V2-V6, II, III, and aVF. Transthoracic echocardiography (TTE) revealed a large pericardial effusion with more than 25% variation of mitral E velocity with respiration and the diastolic collapse of right ventricular outflow tract suggestive of

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cardiac tamponade. No features suggestive of restrictive physiology were detected. Immediate pericardiocentesis of 1.2 L hemorrhagic fluid improved his hemodynamics, but the PND persisted pointing toward possible myocardial disease.

The pericardial fluid analysis revealed an exudative effusion with adenosine deaminase level of 16.8 IU/L, while the smear for acid-fast bacilli and Gene Xpert for tuberculosis were negative. Pericardial fluid cytology showed a discrete population of atypical lymphoid cells, 1.5–2.5 times the size of mature lymphocytes with opened up chromatin, high nuclear: cytoplasmic ratio, and scanty cytoplasm. In immunohistochemistry, cluster of differentiation (CD) 45 positive cells were gated. Of these, 98% were CD 3, CD 5, CD 43, and CD 38 positive. These cells showed dual positivity for CD 4 and CD 8, negative for CD 19, CD 20, CD 34, CD 23, and no light chain reaction was noted. All these features suggested T-cell lymphoblastic lymphoma (T-LBL).

A repeat 2D TTE after complete pericardiocentesis showed bi-atrial enlargement, biventricular wall thickening [Figure 1a], Grade III diastolic dysfunction (septal e' was 5.9 cm/s, E/A was 2.7 and E/e' was 15), and left ventricular ejection fraction of 55% suggestive of RCM [Figures 2a-c], possibly due to infiltration by tumor cells. Inferior vena cava was dilated and noncollapsible. A visible echogenic ring-like encasement of the ascending aorta, descending thoracic aorta, main pulmonary artery, and proximal segment of its branches indicated tumor infiltration [Figures 1b-d, 2d and Supplementary Videos 1 and 2].

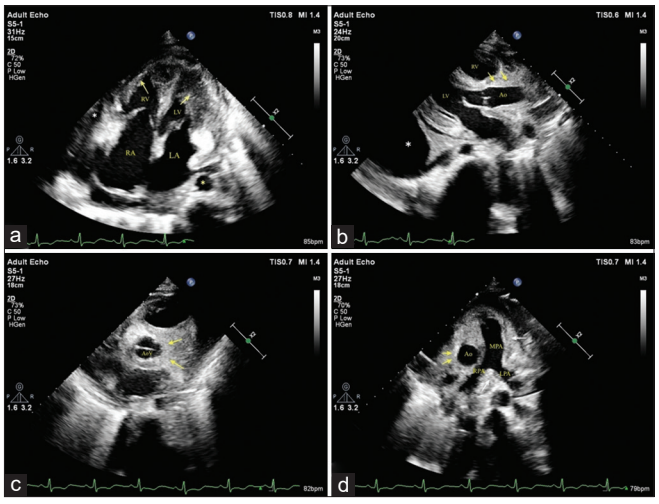

Figure 1: (a) Postpericardiocentesis echocardiography showing mild pericardial effusion (white asterisk), right ventricular and left ventricular (yellow arrow) thickening and thickening of the descending thoracic aorta (yellow asterisk). (b) Pleural effusion (white asterisk), pericardial effusion, and infiltration of aorta (yellow arrows). (c) Lymphoma encasing the aorta. (d) Lymphoma infiltrating aorta (yellow arrows) and the pulmonary artery (white arrows)

Contrast-enhanced computed tomography (CT) scan of the chest [Figure 3] revealed ill-defined sheet-like multi-compartmental soft-tissue lesions in the anterior mediastinum, insinuating between the great vessels and middle mediastinum. There was a pericardial extension of the lesion on the right anterior aspect of the mediastinum with moderate pericardial effusion. The lesion was also seen encasing great vessels. Inferior vena cava and hepatic veins were dilated with reflux of contrast.

Bone marrow examination revealed no involvement. After confirming the diagnosis of T-LBL, he was started on chemotherapy with a modified BFM-2000 (Berlin Frankfurt Munster) protocol with vincristine, daunorubicin, and L-asparaginase along with a corticosteroid. Within 1 week of beginning the chemotherapy, his pericardial effusion resolved, and the pigtail drain was removed. Four days after, the second cycle of L-asparaginase, he developed focal seizures along with right hemiparesis. CT scan of the brain revealed left frontal lobe infarct with hemorrhagic transformation and subarachnoid hemorrhage suggestive of possible cerebral venous thrombosis. CT venography and angiography of intracranial vessels were unremarkable. Magnetic resonance imaging brain and further digital subtraction angiography of intracranial vessels could not be done. His seizure resolved and limb weakness completely resolved over 2 weeks. Due to a deterioration in his performance status, chemotherapy was de-escalated after discussion with the family. Despite the initial improvement, he succumbed to his illness 8 months later to severe community-acquired pneumonia.

DISCUSSION

Cardiac involvement in T-cell lymphoma is not uncommon.[2] The direct extension can lead to pericardial

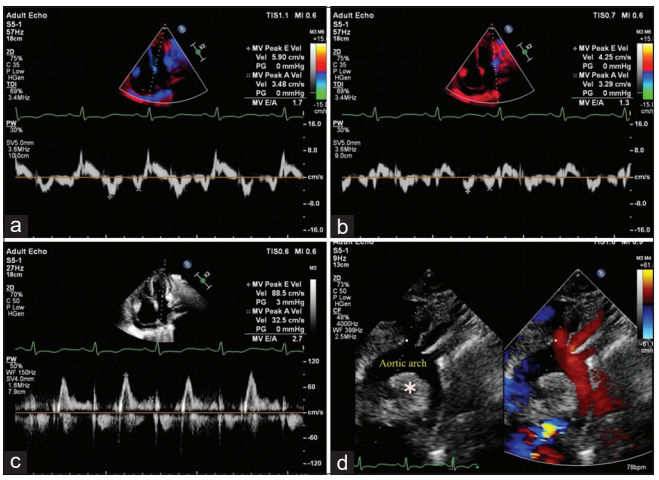

Figure 2: (a) Septal mitral tissue Doppler showing e' of 5.9 cm/s and a' of 3.5 cm/s. (b) Lateral mitral tissue Doppler showing e' of 4.25 cm/s and a' of 3.3 cm/s. (c) Mitral inflow Doppler showing E of 88.5 cm/s and A of 32.5 cm/s (E/A of 2.7). (d) Suprasternal view showing tumor mass between the aortic arch and pulmonary artery (asterisk)

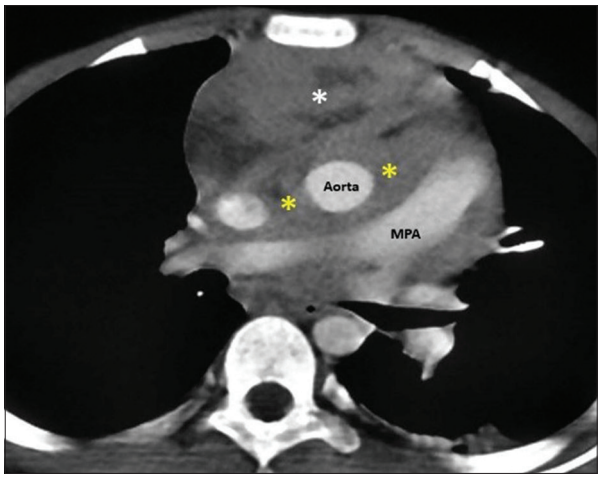

Figure 3: Computed tomography showing ill-defined sheet-like multi-compartmental soft-tissue lesions in the anterior mediastinum (white asterisks) and insinuating between the great vessels and middle mediastinum (yellow asterisk). MPA: Main pulmonary artery

effusion and compression of cardiac structures. In patients with lymphoma, dyspnea related to diastolic heart failure is scarcely reported in the literature.[4-7] The presence of pulmonary venous hypertension in chest X-ray and elevated NT pro-BNP support the diagnosis of heart failure in the index case. Although his hemodynamics improved, the persistence of PND after pericardiocentesis in our patient supports its origin to diastolic dysfunction because of RCM rather than cardiac tamponade. Pericardial effusion is the commonest echocardiographic feature of cardiac involvement in patients of NHL.[8] Massive pericardial effusion and cardiac tamponade as a primary manifestation of lymphoma is rare and mainly occurs in B-cell lymphoma.[3] Literature review reveals only a few published reports of T-cell lymphoma with cardiac tamponade.[9] Although dyspnea is common to both pericardial and myocardial involvement, associated orthopnea or PND as in our case is not specific for either of these. As it is difficult to differentiate based on presenting symptoms alone, a detailed echocardiography reassessment is warranted after the pericardiocentesis to help differentiate between these entities. The occurrence of RCM with lymphomas is extremely rare, and its evidence is limited to a few case reports.[4-6] Earlier studies have indicated toward a higher incidence of cardiomyopathy in patients with lymphoma.[10] Our patient developed cerebrovascular accident likely related to L-asparaginase therapy which is known to significantly increase the risk of thrombotic events (5.2%). After recovering from the cerebrovascular accident, he continued to receive nonasparaginase-based chemotherapy.

The index case illustrates that T-LBL can have an atypical presentation with cardiac tamponade and RCM, requiring careful reassessment after pericardiocentesis. It is the first report of these rare coexisting cardiac pathologies in a patient with T-cell lymphoma to the best of our knowledge. Although the nonavailability of CMR limits our case, the pattern of myocardial involvement points toward tumor infiltration and warrants evaluation for the underlying cause.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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