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Case Report

CD20 negative B-cell lymphoma presenting as constrictive pericarditis: Rare etiology of acute heart failure

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ABSTRACT

We describe a case report of a young girl, who presented with symptoms and signs of acute decompensated heart failure not responding to decongestive therapy. Routine 2D echocardiography revealed a diagnosis of constrictive pericarditis. She underwent pericardiectomy, and etiology of it turned out to be CD20 negative B-cell lymphoblastic primary cardiac lymphoma (PCL). Despite intensive medical and surgical management, it was fatal. PCL is uncommon among cardiac tumors and extremely rare in immunocompetent patients. Constrictive pericarditis is a rare clinical diagnosis and manifestation of pericardial involvement in PCL.

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1. Introduction

Constrictive pericarditis has become a rare manifestation of the pericardial diseases, as most common etiology was tuberculosis in India. Still we encounter sporadic cases of constrictive physiology secondary to rare etiologies like drug-resistant tuberculosis, malignancy, post cardiac surgery or radiotherapy, etc. We report a case of CD20 negative B-cell cardiac lymphoma presented as a diastolic heart failure secondary to infiltration of the pericardium primarily with malignant cells leading to constriction.

2. Case report

A 17-year-old girl presented with gradual onset breathlessness, initially on exertion, NYHA class II since 2 weeks which progressed to NYHA class IV. She had an episode of febrile illness and chest X-ray showed right plural effusion, 6 months ago. Pleural fluid analysis was suggestive of tuberculous etiology; hence, anti-tuberculous treatment was started. In the emergency room, she was tachypneic at rest and room air saturation was 85%. Clinically, there was sinus tachycardia with heart rate of 124 min⁻¹. On auscultation, there was no

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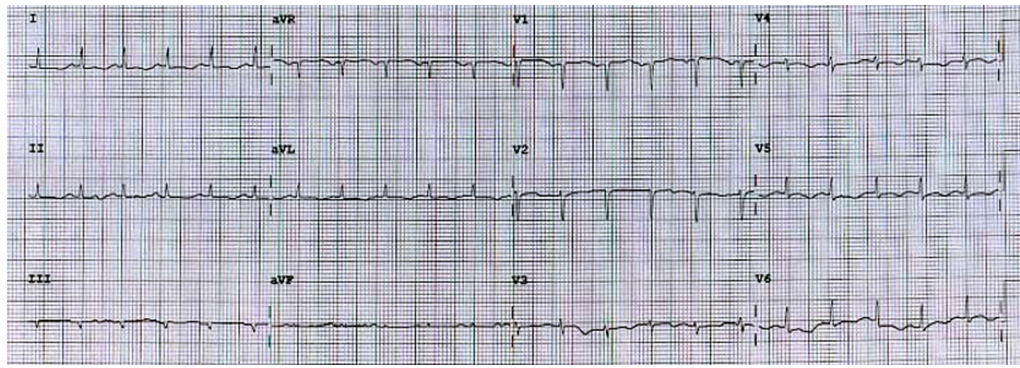


Fig. 1 – Electrocardiogram showing sinus tachycardia with low voltage complexes particularly of limb leads. There are no significant ST-T changes.

murmur, but decreased breath sounds bilaterally in both lower one third of the chest. Chest X-ray showed mild cardiomegaly with bilateral pleural effusion. ECG showed sinus tachycardia with low voltage complexes (Fig 1). The differential diagnosis of disseminated multi-drug-resistant tuberculosis, high-grade lymphoma and collagen vascular disease was made. Echocardiography was suggestive of effusive constrictive pericarditis such as >50% variations of Doppler velocities, thickened pericardium, dilated inferior vena cava, noncollapsing with respiration (Figs. 2A–D and 3A–D). Also there was evidence of minimal ascites and bilateral mild pleural effusion on sonographic examination of abdomen and chest. Laboratory investigations revealed mild anemia [Hb – 8.9 g%], a slight increase in TLC with lymphocyte predominance with normal

ESR initially. Peripheral smear showed normocytic normochromic red blood cells, absolute lymphocytic leukocytosis, and adequate platelets. Liver function test showed mild elevation of bilirubin, liver enzymes, and mild hypoalbuminaemia. Serological markers (such as antinuclear antibody and RA factor) of collagen vascular diseases were negative. Serum LDH level was 652 U/L that is usually elevated in lymphomas. Pleural effusion fluid analysis showed predominance of lymphocytes with no evidence of any organism. ADA level was 72 mg/dl. Pleural fluid cytology showed predominantly lymphocytes, few neutrophils, and reactive mesothelial cells. There were no malignant cells. Her viral screening including HIV-1 and HIV-2 was nonreactive. Laboratory investigations were not conclusive of any possible etiology

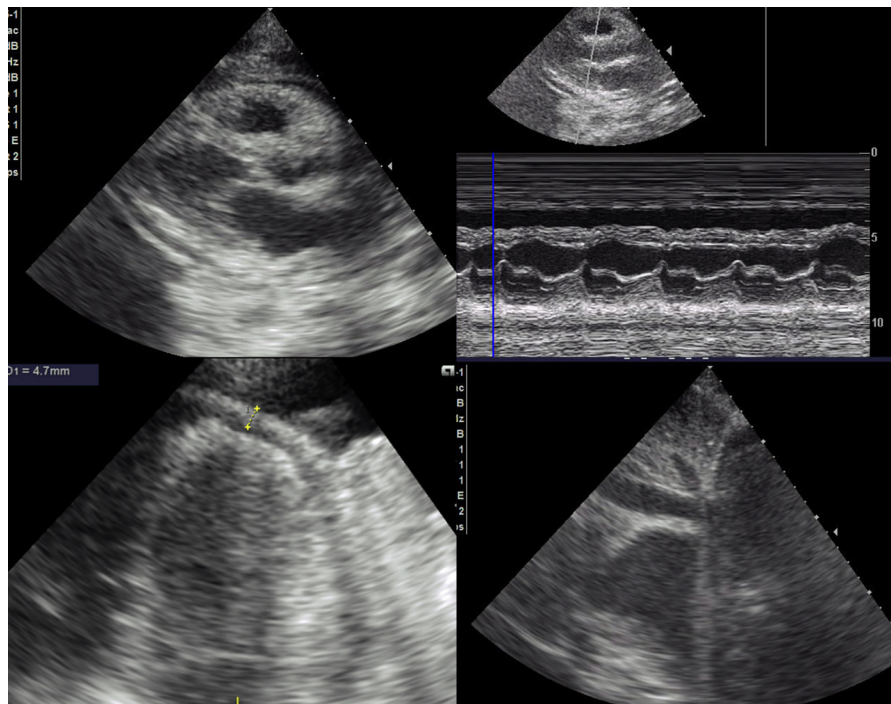


Fig. 2 – (A–D) 2D echocardiography showing thickened pericardium with mild circumferential pericardial effusion in parasternal long axis view (A); M-mode echocardiography revealed septal bounce (arrow) with thickened pericardium and mild pericardial effusion (B). Zoomed 4 chamber view (C) showed thickened pericardium (4.7 mm) and subcostal view (D) showing dilated inferior vena cava non-collapsing with respiration.

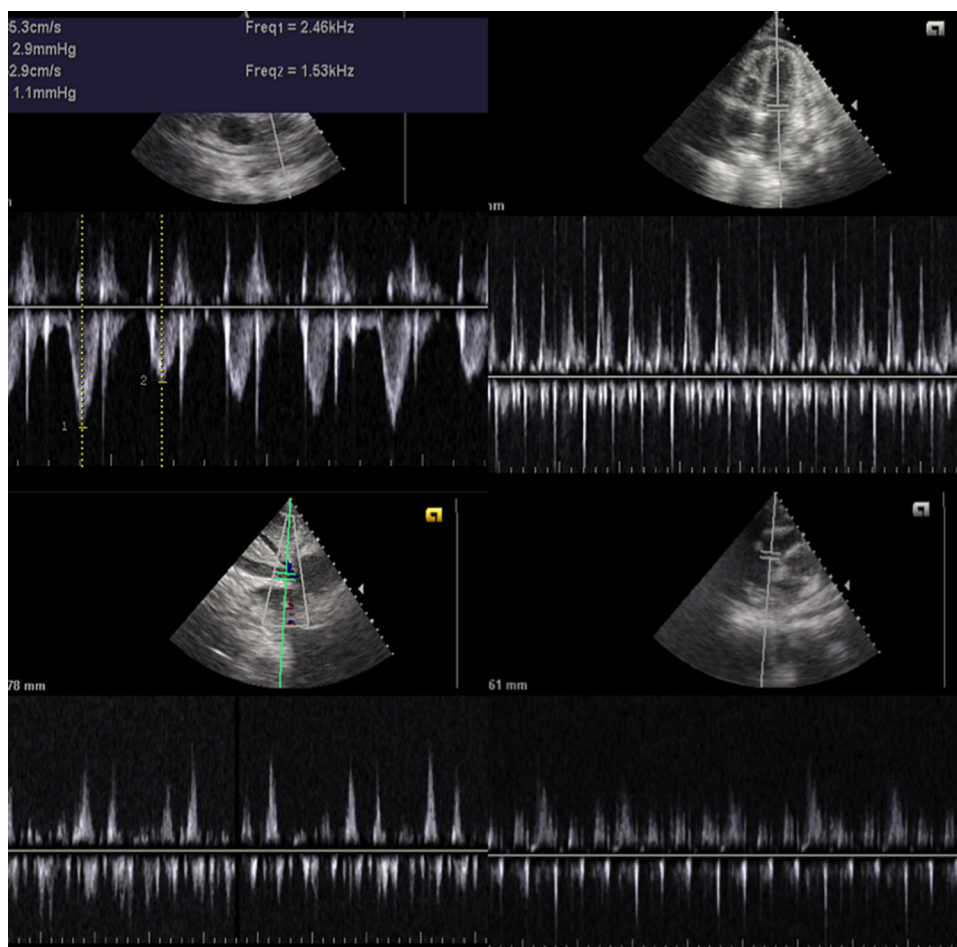


Fig. 3 – (A–D) Doppler velocities across the pulmonary artery showed >50% Doppler variation with respiration (A). Also there was >50% Doppler variation with respiration of velocities of mitral and tricuspid valve (B and C) suggestive of ventricular interdependence. Hepatic vein Doppler also showed systolic flow reversal (D).

of the cause of constriction. Hence, a possible diagnosis of tuberculous constrictive pericarditis was made. Pleural fluid was negative for any bacterial growth after 48 h of incubation.

Contrast-enhanced CT scans of chest revealed soft tissue density mass encasing cardiac chambers with significant bilateral pleural effusion with passive collapse of both lungs.

Meanwhile, there was no response to conservative anti-failure treatment hence, the case was discussed with cardiothoracic surgeon and planned for surgical pericardiectomy. She underwent total pericardiectomy through midline thoracotomy, pericardium was grossly thickened, adherent to sternum and myocardium (Fig. 4A–D). There were few areas of fibrosis and calcifications. Post operatively, the patient improved significantly with reduction of her symptoms.

Histopathological examination of pericardial biopsy report showed fibro-adipose tissue of pericardium and adjacent striated muscles infiltrated with monomorphic lymphocytes with high mitotic activity. These infiltrating cells were positive for CD45 [LCA], negative for CD3, CD20, and CD138, weak-to-moderate positive for terminal deoxynucleotidyl transferase (TdT), high MIB-1 index [70–80%]. Chemotherapy in consultation

with medical oncologist was started in the form of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP regime).

After initial improvement, her symptoms worsened further 1 week later in the form of progressive breathlessness, requiring noninvasive ventilation. She developed type II respiratory failure hence, electively intubated and ventilated. She had persistent hypotension and hyponatremia, which required high doses of inotropes, intravenous sodium correction. Despite our best efforts, she died secondary to multi-organ dysfunction.

3. Methods and materials

We searched Google, PUBMED, MEDLINE, EMBASE using keyword like 'Acute decompensated Heart failure, Pericardial effusion, Constrictive Pericarditis, Primary cardiac lymphoma'. Then refined search for cardiac lymphomas presenting as constrictive pericarditis was done. After evaluating the cross references of earlier articles, we searched for the CD20 negative lymphomas. Among all the articles we tried

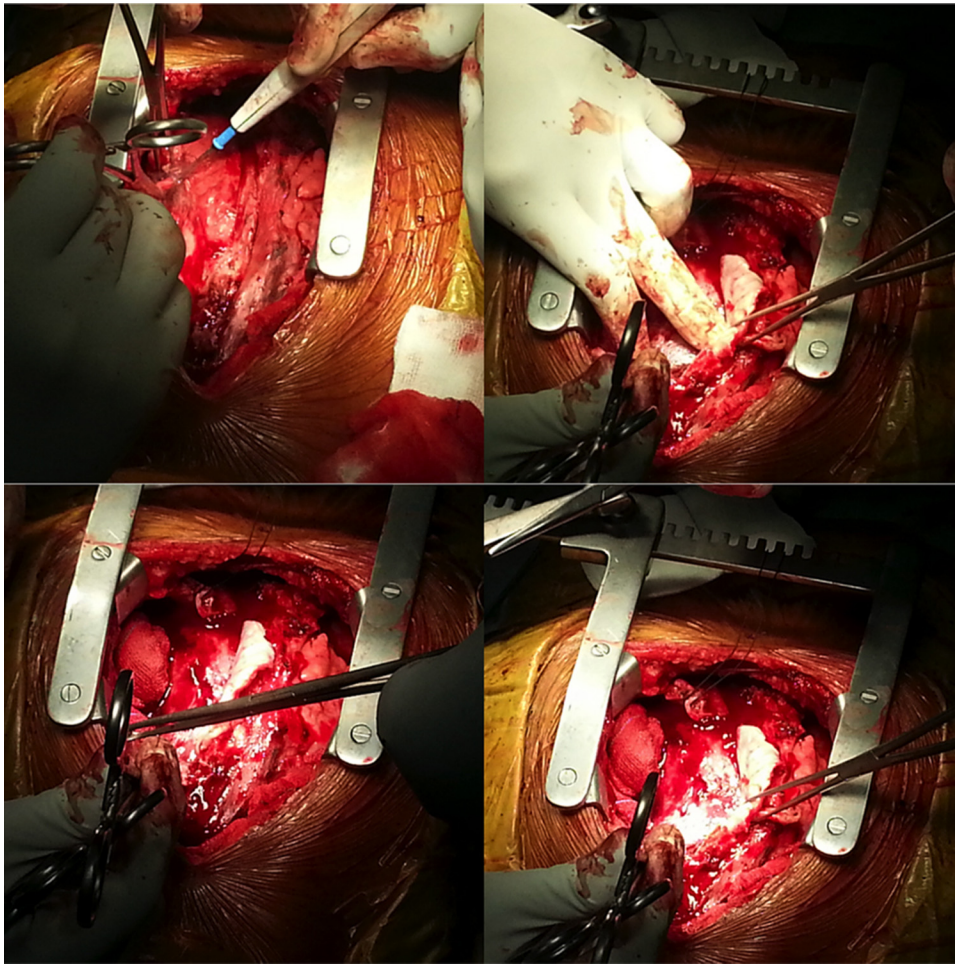


Fig. 4 - (A-D) Midline sternotomy revealed thickened pericardium (A), which was dissected using electro cautery and blunt dissection using fingers (B and C). Pericardium was thickened and firmly adherent to underlying epicardium with areas of fibrosis and calcifications (D).

correlating our clinical features and our case was unique that it was rare combinations of all the features.

4. Discussion

Constrictive pericarditis is a rare presentation of primary cardiac lymphoma (PCL) involving pericardium. PCL is defined as a non-Hodgkin's lymphoma (NHL) involving only the heart and/or pericardium or as an NHL with the bulk of the tumor located on the heart.¹ PCLs are extremely rare cardiac tumors in immunocompetent patients. It is estimated that their incidence is <1% of all cardiac tumors.^{2,3} Usually, most of the cases are of NHL in immunocompromised patient, particularly in patients with acquired immunodeficiency syndrome.² It involves mostly right-sided structures, particularly right atrium in 66% of cases. Overall pericardial involvement is estimated to be present in 30–58% of these cases of PCL, manifested as isolated pericardial effusions (12%) or in combination with mass lesions (44%).^{1,4} Primary involvement of pericardium was seen in 16% of patients and presentation as a constrictive pericarditis is very rare.¹

The most frequent cardiac clinical manifestations associated with PCL are recurrent pericardial effusion, massive pericardial effusion with cardiac tamponade, systolic and diastolic heart failure, and atrioventricular block (AV-block).^{5,6} However, cytologic examination of cardiac tumor or pericardial effusion is paramount for a definite diagnosis of this condition. Prognosis of PCL is poor with a median survival of 7 months due to diagnostic delay, and usually it is a fatal condition despite treatment as in our case.^{1,7}

Ho et al.³ also described similar case like ours of PCL in a immunocompetent patient presenting as a constrictive pericarditis. Venance et al.⁸ reported a case of young man similar to our case who presented with constrictive physiology following infiltration of pericardium with CD30 positive T cell type. Both the cases described by authors had fatal outcome as in our case.

Bertog et al.⁹ presented long-term survival of 163 patients after pericardiectomy in constrictive pericarditis patients. Lower sodium levels were one of the bad prognostic markers apart from the etiology of the constrictive pericarditis. In their series, there were 2 cases of lymphoma presenting as constrictive physiology.

The management for PCL is mainly chemotherapy combined with surgical intervention for large tumors causing mechanical obstruction such as large mass or constrictive physiology.¹⁰ Umakanthan et al. reported a case of precursor B-cell lymphoblastic PCL, where CD20 expression was negative as in our case. CD20 antigen is a membrane-bound protein and marker of normal and neoplastic B lymphocytes. A total of 90–95% of diffuse large B-cell lymphomas, which are common type of lymphomas expresses it and helps in selecting chemotherapeutic regimen like addition of rituximab is directed against CD20. CD20 is not expressed on immature B precursors and plasma cells.^{11,12} Among all the types of B-cell lymphomas, the incidence of CD20 negative lymphomas was 3% (232/7) in the series of Gaur et al. Of the 7 cases, 5 cases were of plasmablastic lymphomas, while two were unclassifiable B-cell lymphomas. Plasma cell markers are CD38, CD138. In our case, CD138 was also negative. Prognosis is usually dismal for CD20 negative lymphomas as response to chemotherapy is poor. Our case report is first in the literature, where CD20 negative B-cell PCL presented as constrictive pericarditis.

5. Conclusion

PCLs are rare among cardiac tumors and still an uncommon presentation of pericardial involvement in immunocompetent patient. But these patients have poor outcome despite treatment.

Conflicts of interest

The authors have none to declare.

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