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Case Report

Primary Mediastinal T-Cell Non-Hodgkin's Lymphoma Presenting as Cardiac Temponade: a Diagnostic Dilemma

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ABSTRACT

A T-cell lymphoblastic lymphoma is an uncommon subtype of Non-Hodgkin's lymphoma which can rarely present with massive pericardial effusion or cardiac temponade. We report a case of T-LBL in a 20 year old female who presented with features of cardiac temponade. Contrast enhanced computed tomography (CECT) scan of the thorax showed presence of a mediastinal mass. Cytological examination of pericardial fluid and needle aspirate of the mass showed features consistent with a lymphoblastic lymphoma, which was later confirmed as T-cell lymphoblastic lymphoma on histological and immunohistological examination. Mediastinal T-cell lymphoblastic lymphoma presents a diagnostic dilemma and must be distinguished from other closely resembling mediastinal tumors presenting with cardiac temponade by histological and immunohistological examination. Also, the cytological examination of pericardial fluid and aspiration cytology can be used as reliable tool in diagnosing lymphoblastic lymphoma, aiding in further evaluation of the patient with the advantage of early management and intervention.

Keywords: Lymphoblastic; Mediastinal; Pericardial; Temponade

INTRODUCTION

A T-cell lymphoblastic lymphoma (T-LBL) is an uncommon subtype of adult Non-Hodgkin's lymphoma with an incidence of only 2%. [(1)] T-LBL may presents as mediastinal mass, but its presentation as cardiac temponade is rare. [(2)] Malignant pericardial effusion associated with T-LBL is a sign of advanced malignancy and poor prognosis. [(3)]

CASE PRESENTATION

A 20-yr-old Indian female presented with the complaint of progressive shortness of breath, chest pain and fatigue for the past 1 month. There was no history of fever, chills, loss of appetite, significant weight loss, drug intake (immuno-suppressive drugs) or any auto-immune or chronic systemic disease. Her family and past history were also unremarkable. On general examination, patient was of average built, mildly anemic and tachypnic. There was no cervical, axillary or inguinal

lymphadenopathy or any organomegaly. Her pulse was 110/min and B.P was 80/60 which showed pulsus paradoxus. Auscultation of the chest revealed clear lungs and muffled heart sounds with frictional rub. Chest X-ray showed increased cardiac silhouette. Electrocardiography (ECG) revealed diffuse ST elevation in association with PR depression and electrical alternans in the QRS wave, with beat-to-beat variation in the direction and amplitude, suggestive of cardiac temponade. Routine hematological investigations, bone marrow and CSF examination were unremarkable. Her serum electrolytes levels, serum adenosine deaminase (ADA) level, coagulation profile, thyroid, liver and renal function tests, were within normal limits. However, serum lactic acid dehydrogenase (LDH) level was markedly raised to 1000 U/L (Reference range: 115-221 U/L). Contrast enhanced computed tomography (CECT) scan of the thorax showed hypodense collection in pericardial space, suggestive of massive pericardial effusion (Figure 1a) and a homogeneously enhancing soft tissue density mass lesion measuring 8 x 9 x 8 cm involving the anterior and superior mediastinum (Figure 1b). The mass was seen extending from thoracic inlet to the level of superior pulmonary vein inferiorly, displacing the left brachiocephalic vein posteriorly. Cardiac chambers and vessels appeared normal. The radiological findings were suggestive of a mediastinal mass probably arising from thymus resulting in cardiac temponade. Pericardiocentesis was immediately done to relieve the immediate worsening symptoms. About 1000 ml of pericardial fluid was aspirated and was sent for microbiological, biochemical and cytological examination. Gram staining and acid fast bacilli (AFB) staining and culture of the pericardial fluid were negative for bacteria. However, the cytological examination of pericardial fluid (Figures 1c&1d) and fine needle aspirate from the mediastinal mass showed predominantly immature intermediate sized cells having high nucleo-cytoplasmic ratio (N:C), cleaved or convoluted nucleus with finely dispersed granular chromatin, inconspicuous nucleoli and scant cytoplasm, along with mature lymphocytes (Figures 2a).

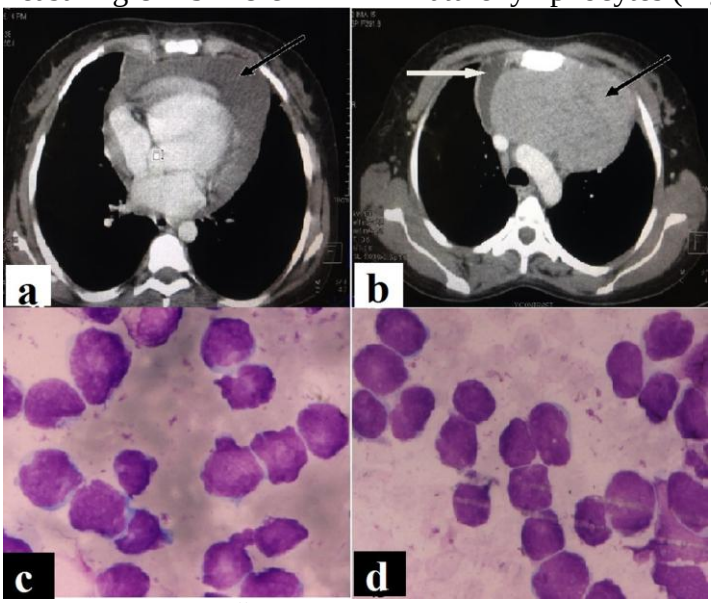

Figure 1: a) CECT image showing massive pericardial effusion (arrow) resulting in cardiac temponade, b) showing homogenously enhancing mediastinal mass (black arrow) with pericardial effusion (white arrow), c) and d) Pericardial fluid cytology showing lymphoblasts having high N:C ratio, cleaved or convoluted nucleus with finely dispersed granular chromatin, inconspicuous nucleoli and scant cytoplasm (May-Grunnwald Giemsa stain x 500)

Based on cytological findings, a diagnosis of a Non-Hodgkin's lymphoma favouring a lymphoblastic lymphoma was made. Histological examination of the biopsy obtained also confirmed the cytological diagnosis of lymphoblastic lymphoma (Figure 2b). Immunohisto-chemical examination further revealed tumor cells showing membranous positivity to T-cell immunomarkers such as CD-3 (Figure 2d), CD-7 and CD-2. The tumor cells also showed nuclear positivity to immaturity marker Terminal deoxy - nucleotidyl (Tdt) (Figure 2c), indicating their lymphoblastic origin. Further, B-cell markers such as CD-19, CD-20, CD-79a, bcl-6, multiple myeloma oncogene (MUM-1) were found to be negative. Also, tumor cells showed non-reactivity to CD 34, CD-5, CD-10, CD-30, CD-15, CD-23, CD-56, cytokeratin and neuroendocrine immunomarker synaptophysin. Based on radiological, cytological and immunohistochemical findings, a diagnosis of mediastinal T-cell lymphoblastic lymphoma presenting as cardiac temponade was made.

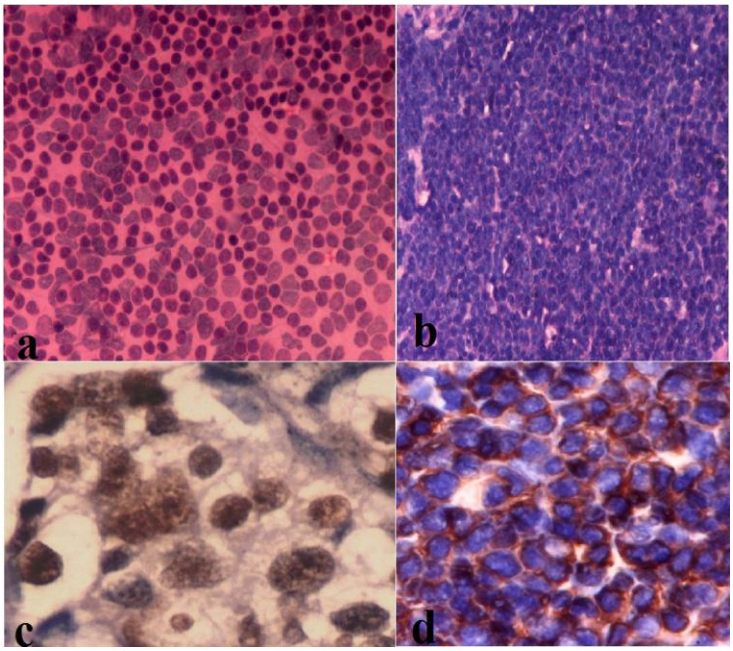

Figure 2: a) Aspirate cytology smear of mediastinal mass showing lymphoblasts admixed with lymphocytes, suggestive of lymphoblastic lymphoma (H&E x 500), b) histological section showing predominantly lymphoblasts (H&E x 125), c) and d) showing nuclear positivity of tumor cells to Terminal deoxy-nucleotidyl (Tdt) and membranous CD-3 positivity respectively, confirming the T-cell origin (c x 1000, d x 1000).

DISCUSSION

Mediastinal T-cell lymphoblastic lymphoma presenting as cardiac temponade pose a diagnostic challenge as the clinical manifestations of the disease are wide and variable and may overlap with other diseases. A mediastinal tumor or mass may sometimes be clinically silent and may not produce obvious signs and symptoms. Though disseminated systemic lymphoma often has involvement of the mediastinum but only 5% to 10% of patients with lymphoma present with primary mediastinal lesions. [(4)] Pericardial effusion may sometimes be the only presentation with underlying mediastinal mass or tumor, only detectable incidentally on radiological imaging. Therefore, patient with massive pericardial effusion should always be investigated for any occult malignancy.

Diagnosis of T-LBL is made when a large nodal/extranodal tumor mass is present with lymphoblast count of less than 25% in the bone marrow. In the present case, there was no evidence of leukemia or bone marrow involvement or any disseminated lymphoma but the cytological examination of the pericardial fluid revealed lymphoblastic cells, raising suspicion for underlying malignant mass which was radiologically evident on CECT.

The differential diagnosis in the present case included other tumors presenting as mediastinal mass including thymoma and thymic carcinoma, Hodgkin's lymphoma (nodular sclerosis subtype), primary mediastinal B-cell lymphoma with sclerosis, Burkitt lymphoma, Extranodal NK/T-cell lymphoma, blastoid variant of mantle cell lymphoma and germ cell tumors.

Malignancies of the thymus such as thymoma and thymic carcinoma may cause diagnostic confusion with T-LBL arising from thymus. [(5)] However, in the present case, the cytomorphology of immature cells in pericardial fluid and fine needle aspirate from the mass was lymphoblastic which was further confirmed by positive immune-expression of tumor cells to T-cell markers such as CD-3 and CD-7 and non-reactivity to cytokeratin-19, ruling out epithelial origin of the tumor.

Hodgkin's lymphoma, particularly the nodular sclerosis subtype may also present as mediastinal mass producing malignant pericardial effusion and temponade but is distinguishable from T-LBL by its distinct histomorphology and CD-30 and CD-15 immuno-expression of the Reed Sternberg cells. [(3)]

Primary mediastinal B-cell lymphoma with sclerosis (PMBL) is a subtype of the diffuse large B cell lymphoma, frequently occurring as a mediastinal neoplasm. [(6)] It is differentiable from T-LBL by positive B-cell immune-expression such as CD-19, CD-20, CD-79a and non-reactivity to T-cell immunomarkers CD-3 and CD-7 by its tumor cells. [(6)]

Burkitt lymphoma may rarely present with primary mediastinal mass associated with cardiac temponade. [(7)] Cytologically the cells are of intermediate size and show coarse chromatin in contrast to lymphoblastic lymphoma cells having fine granular chromatin. Also, the cells express B-cell immunophenotype. [(7)]

Extranodal NK/T-cell lymphoma may rarely involve the pericardium but it was excluded as the cells in the present case showed non-reactivity to CD-56. [(8)] Though, neoplastic cells of blastoid variant of mantle cell lymphoma (B-MCL) can resemble T-LBL but it lacks CD-23 immuno-expression and show characteristic CD-5 positivity. [(9)]

Effusions in lymphoblastic lymphoma and other high grade lymphomas are associated with poor outcome. For remission induction, patient in the present case was kept on CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) regime. Considerable symptomatic improvement was noticed after first cycle of chemotherapy. Consolidation and maintenance therapy are also planned in future follow-up.

CONCLUSION

T-LBL can rarely present as cardiac temponade and pose a diagnostic challenge. Patients presenting with cardiac temponade should always be investigated for underlying occult malignancy. A T-LBL must be differentiated from other closely simulating lymphomas and mediastinal tumors presenting as cardiac temponade, by histological and immunohistochemical examination. The findings in the present case also underlines the utility of cytological examination of pericardial fluid and aspiration cytology of the mediastinal mass, which can be used as reliable tool in

diagnosing lymphoblastic lymphoma, aiding in further evaluation of the patient with the advantage of early management and intervention.

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