

**Case Report**

T Cell Lymphoma: A Rough Road to Diagnosis

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Abstract

Introduction: Lymphoproliferative disorders are well known for their variable presentation. Peripheral T cell lymphoma not otherwise specified (PTCL-NOS) is one subtype which is especially notorious. It has a variable age of onset, predominantly a nodal presentation and immunohistochemistry (IHC) depicting mature T cells.

Presentation of case: We share our experience of a 33 year old patient who presented with mediastinal mass with pericardial effusion. However, the histopathology revealed PTCL-NOS.

Conclusion: This case is one of the manifestations of heterogeneity encompassing T cell neoplasms where discordance in clinical and pathological behavior caused not only diagnostic confusion but also complicated treatment decisions.

Keywords: lymphoma; Mediastinum; T cell

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Consent: Consent was taken from the patient for publication of this case report.

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Introduction

Peripheral T cell lymphoma not otherwise specified (PTCL-NOS) is the most common T cell neoplasm. Though, there is growing emphasis on classification and grouping based on immunohistological features, PTCL-NOS still remains an umbrella entity which shows heterogeneity in presentation and prognosis. Here we present a patient who had clinical features mimicking T cell lymphoblastic lymphoma, only to reveal pathological diagnosis of PTCL-NOS in the background of several unusual histological features.

Case summary

A thirty three year old non-smoker male presented with dry cough, progressive shortness of breath and right side pleuritic chest pain for three months. He had significant loss of weight and appetite. There was no history of fever, glandular swelling, night sweats or haemoptysis. He was a diabetic with adequately controlled blood sugar level on oral hypoglycaemic agents. Chest imaging by Contrast enhanced computed tomography (CECT) revealed bilateral pleural effusion (right more than left), pericardial effusion and anterior mediastinal mass of size 6X 3.5 cm with heterogeneous enhancement (figure 1a and figure 1b). Multiple enlarged pretracheal, paratracheal, prevascular lymph nodes were noted. Other investigations revealed neutrophilic leucocytosis with normal haemoglobin and platelet count. His liver function and kidney function tests were normal. HIV serology was negative and serum lactate dehydrogenase level was 1.5 times the upper limit of normal.

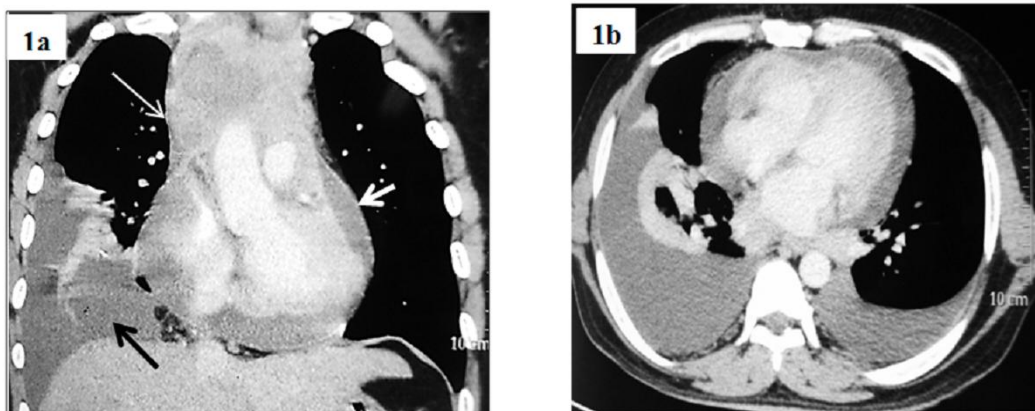


Figure 1a: Coronal section to show mediastinal mass (thin white arrow), right pleural effusion (black arrow) and pericardial effusion (thick white). **Figure 1b:** Axial CECT images depict bilateral pleural and pericardial effusion

An open mediastinal biopsy was performed. It revealed malignant round cell tumour with extensive fibrosis. Presence of extensive fibrosis lead to inadequacy of tissue sampling on image guided attempts to biopsy hence necessitating open biopsy. On histopathological evaluation tumour cells were intermediate in size with scant cytoplasm (figure 2a). They stained immunopositive for CD3, CD5, CD7 diffusely and focally for epithelial membrane antigen and CD30 (figure 2b-2c). Stains for Mum 1, CD20, Alk-1, Bcl-16, CD34 and TdT were negative (figure 2d-2f). Very high proliferation index with Ki67 greater than 80% was

found. Bone marrow examination, testicular ultrasound, spinal fluid analysis and CECT abdomen were normal.

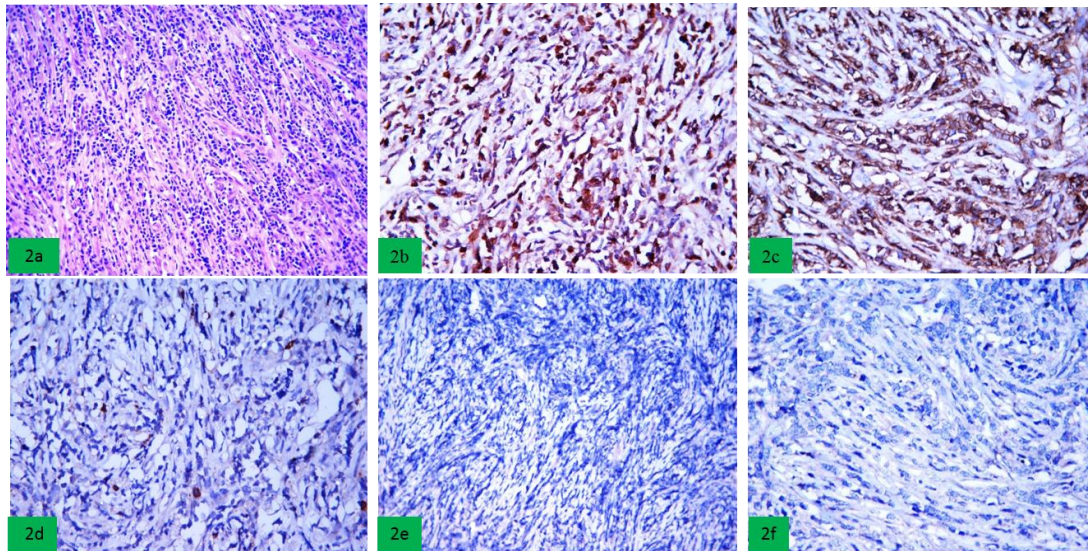


Figure 2a. Low power photomicrograph shows a malignant round cell tumor arranged in cords with extensive desmoplasia (X20); On immunohistochemistry, tumor cells are immunoreactive with CD3(2b),CD7(2c) while they are immunonegative for CD20 (2d), TdT(2e) and ALK1(2f)

In view of clinical behaviour coupled with high mitotic index, treatment based on the lines of aggressive lymphoma protocols was initiated. Post induction the mediastinal mass has significantly decreased in size with resolution of pleural and pericardial effusion. The patient has currently completed consolidation chemotherapy and continues to be in remission.

Discussion

Differential diagnosis for mediastinal mass with pericardial and pleural effusion in this age group includes germ cell tumour, lymphoproliferative disorder and lung carcinoma. The histological picture pointed toward T cell lymphoproliferative disorder though several atypical features were observed.

The presence of fibrosis in a T cell tumour is a rare occurrence. It is a feature of primary mediastinal B cell lymphoma [1] and Hodgkin's lymphoma especially the nodular sclerosis variety [2]. In T cell lymphomas, it is seen in anaplastic large cell lymphoma [3] and has been reported in cutaneous T cell lymphoma [4, 5]. Fibrosis in lymphoma usually occurs after treatment and is a feature of response to the therapy. However, this one is the first case where a T cell lymphoma with predominant mediastinal involvement and no prior treatment showed significant degree of fibrosis.

The clinical presentation of mediastinal mass along with pericardial effusion and pleural effusion is usually seen in T cell lymphoblastic lymphoma, though the age of our patient is higher than the usual age of presentation. However the IHC picture contradicts a diagnosis of lymphoblastic lymphoma and favours the diagnosis of PTCL-NOS. TdT positivity is hallmark of lymphoblastic lymphoma as it is expressed in more than 85 % of cases [6, 7]. There are only few case reports where TdT has been found to be negative. In those, the diagnosis of lymphoblastic lymphoma has been made by detection of other lymphoblastic or precursor cell markers e.g. CD34, KIT, and CD99 [8]. In our case these markers were found to be negative. This

discordance between clinical and pathological findings complicated the picture. Not only it added to the diagnostic confusion but also interfered in deciding proper line of treatment and prognosis.

PTCL-NOS present with both extranodal and nodal involvement in upto 50 percent cases [9]. The most common extranodal sites to be involved are skin and gastrointestinal tract. However, mediastinal involvement with pleural effusion and pericardial effusion has been seen in only few cases and is associated with shorter progression free survival [10].

Conclusion

Lymphomas are known for variations in biological behaviour and clinical presentations. This case is a typical example of difficulties encountered in reaching conclusive diagnosis even in cases where the clinical behaviour certainly points towards one. It remains to be seen if there is difference in response to treatment as well.

Consent

Consent was taken from the patient for publication of this case report.

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