

The sandwiched pleural effusion

Sir,

T-cell lymphoblastic lymphoma (T-LBL) is a rare type of non-Hodgkin's lymphoma (NHL). Predominantly seen in adolescents and young adults, it usually presents with a mediastinal mass with pleural effusion. Pleural fluid analysis may be misleading in a tuberculosis-endemic country causing a significant delay in diagnosis. A high index of suspicion with some characteristic features on radiology may point towards the diagnosis which can be established by thoracoscopic pleural biopsy.

A 60-year-old gentleman presented with gradually progressive dyspnea, left-sided chest pain, and weight loss for 2 months. There was no history of cough, fever, or night sweats. He was a nonsmoker with no comorbidities, past environmental/occupational exposures, or tuberculosis. He had been started on empirical antitubercular therapy by his physician with no symptomatic relief. Respiratory examination revealed signs of left pleural effusion.

Basic laboratory investigations were normal, and HIV serology was negative. Chest radiograph revealed massive left pleural effusion, and there was nodular pleural thickening on ultrasonography. Pleurocentesis revealed hemorrhagic, exudative, lymphocyte predominant fluid with negative malignant cytology (9354 cells/cu.mm – 98% lymphocytes, protein – 4.19 g/dL, lactate dehydrogenase – 185U/L, glucose – 84 mg/dL, adenosine deaminase – 84.4U/L). On pleuroscopy, large parietal pleural nodules coalescent on the diaphragmatic surface were visualized and biopsied [Figure 1].

Computed tomography (CT) of the thorax revealed loculated left pleural effusion with circumferential pleural thickening and underlying lung collapse; intercostal vessels were encased by the thickened pleura – “the pleural sandwich sign” [Figure 2].

Histopathology revealed fibroconnective tissue diffusely infiltrated by sheets of medium-sized atypical lymphoid cells with moderate cytoplasm, large nuclei, and coarse chromatin. The cells were positive for CD3, B-cell lymphoma (BCL), CD7, leukocyte common antigen, terminal deoxynucleotidyl transferase, and CD99 and negative for CD20, CD23, CD10, CD4, CD8, CD30, and CK. The proliferation index was 70% [Figure 3].

Whole-body positron emission tomography (PET)-CT scan showed metabolically active diffuse left pleural nodular thickening and fluorodeoxyglucose (FDG)-avid left hilar,



Figure 1: Pleuroscopy image showing large parietal pleural nodules

tracheobronchial, precarinal, prevascular, and subcarinal nodes (maximum standard uptake value [SUV] – 7.9 and 7.6, respectively); there were no other lymph nodes [Figure 2]. A diagnosis of T-cell lymphoblastic lymphoma with pleural involvement was made. Once the pleural drainage reduced, the intercostal drain was removed after pleurodesis with povidone-iodine. The patient was started on chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone regimen and is on follow-up.

Pleural effusions associated with NHL can develop through various mechanisms, namely (1) enlarged mediastinal lymph nodes obstructing the lymphatic drainage of the pleura, (2) obstruction of the thoracic duct, and (3) direct involvement of the pleura.^[1]

The most common type of lymphoma reported to involve the pleura is diffuse large BCL, followed by follicular lymphoma.^[2] Lymphoblastic lymphomas are rare accounting for 8% of all lymphomas. T-LBL usually presents at a young age (median – 25 years) with mediastinal mass (90%), pleural effusion (20%), and bone marrow infiltration (25%).^[1,3] T-LBL primarily presenting with extensive pleural involvement is extremely rare.^[4] Predominant pleural involvement with few mediastinal lymph nodes noted on PET-CT with no evidence of any mediastinal mass lesion in an elderly male was unique in our patient.

Pleural fluid analysis can sometimes be misleading due to lymphocyte predominance and high ADA levels, leading to inappropriate antitubercular therapy and a delay in the

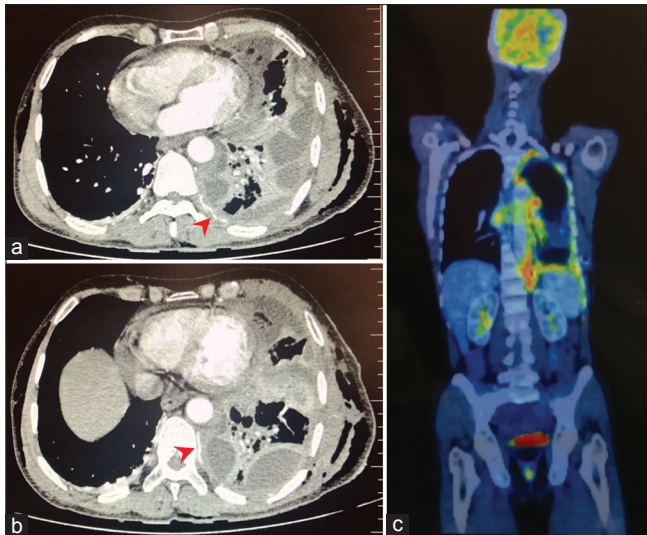


Figure 2: (a and b) Contrast computed tomography chest showing circumferentially thickened pleura encasing the intercostal arteries (arrowheads) – pleural sandwich sign; (c) whole-body positron emission tomography-computed tomography scan showing metabolically active diffuse left pleural nodular thickening and fluorodeoxyglucose-avid left hilar, tracheobronchial, prearcinal, prevascular, and subcarinal nodes

diagnosis of lymphoma, especially in endemic countries. Cytological diagnosis of lymphoma may be difficult as effusions are more likely to be vascular and lymphatic obstruction related. In cases with a positive cytology, flow cytometry or immunohistochemistry on cell blocks may help in diagnosis and subtyping.^[5] Thoracoscopy and biopsy from the representative sites seals the diagnosis.

A thoracic CT scan in lymphoma shows either diffuse nodular pleural thickening, a localized mass, or an isolated effusion. These findings are nonspecific and can be seen in malignant mesothelioma or pleural metastasis. The “pleural sandwich sign,” described as encasement of intercostal vessels by the pleural mass, has been reported to be highly suggestive of a lymphoma, as it is not seen in other pleural malignancies due to early infiltration of the vessels and lack of extrapleural component.^[6] This sign is derived from the “mesenteric sandwich sign” and “thoracic sandwich sign” defined in mesenteric and mediastinal lymphomas, respectively.

PET-CT scan is useful in detecting extranodal involvement; however, majority of the T-cell lymphomas exhibit poor FDG uptake with modest SUV values or even false-positive scans.^[4]

Although prognostic factors for T-LBL in adults are not conclusively identified, pleural effusions in lymphoma usually carry a poor prognosis in the younger age group.^[1,7]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

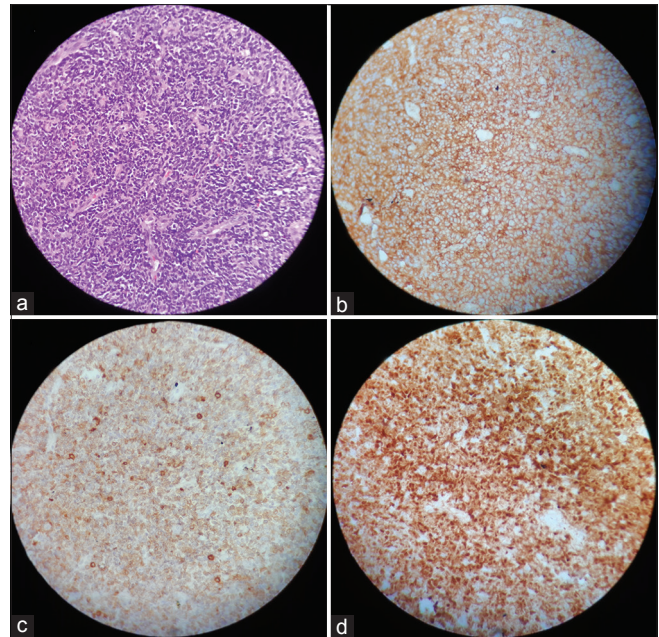


Figure 3: Histopathology of pleural biopsy: (a) Uniform atypical lymphoid cells seen with irregular nuclei and scant cytoplasm (H and E, x400); (b) the neoplastic cells showing positive staining with CD45 immunostain; (c) the neoplastic cells showing weak membranous positivity for CD3 immunostain; (d) the neoplastic cells showing nuclear positivity for terminal deoxynucleotidyl transferase immunostain

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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