Case report: T-Cell Lymphoblastic Lymphoma Presenting as Massive Pleural Effusion

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Abstract

Lymphoblastic lymphoma is the most common type of non-Hodgkin lymphoma (NHL) in children, with T-cell lymphoblastic lymphoma (T-LBL) comprising the majority of cases. It typically presents with an anterior mediastinal mass and disseminated disease. While pleural effusion may occur in up to 30% of T-LBL cases, initial presentation as isolated massive pleural effusion is exceedingly rare and has been described in only a limited number of pediatric cases. Fewer than 15 well-documented reports exist globally, with most describing pleural effusion secondary to a mediastinal mass. We report the case of a seven-year-old boy with T-LBL who initially presented with massive right-sided pleural effusion and mild pericardial effusion. Subsequent evaluation revealed a mediastinal mass. Pleural fluid cytology demonstrated numerous atypical cells. A pan-computed tomography (CT) scan indicated lymphomatous infiltration of the thymus and hilar lymph nodes. Bone marrow analysis showed less than 5% circulating blasts. Cytomorphology and immunophenotyping were consistent with T-LBL. The patient was diagnosed with mediastinal T-cell lymphoblastic lymphoma and was commenced on chemotherapy.

Keywords: Non-Hodgkin lymphoma, T-cell Lymphoma; pleural effusion: Paediatric; Oman

Introduction

T-cell lymphomas are rare and uncommon malignancies in pediatric populations. Diagnosing, treating, and managing T-cell lymphomas has its own unique challenges. This case report describes a child presented with shortness of breath and cough. His Chest X ray showed unilateral massive pleural effusion. Initially managed for suspected infectious etiology, but ultimately diagnosed with T-cell lymphoblastic lymphoma. Early recognition of such cases is crucial for appropriate management.

Case Report

A previously healthy 7-year-old boy presented to the pediatric emergency department at Sultan Qaboos University Hospital with a two-day history of chest pain and lethargy, and one-day history of shortness of breath. There was no reported history of fever, weight loss, night sweats, or bony pain. The child's parents are first-degree cousins, and he has three normal, healthy siblings. There is no family history of malignancy.

Upon general physical examination in the emergency department, the child appeared unwell and pale. He was tachycardiac with a heart rate of 144 beats per minute and tachypnea with a respiratory rate of 36 breaths per minute. His blood pressure and oxygen saturation were within acceptable ranges for his age. There were no palpable cervical lymph nodes felt. Chest examination revealed mild subcostal recession without nasal flaring or grunting. Auscultation of the chest indicated reduced air entry throughout the entire right side, accompanied by fine crepitation and dullness upon percussion. The remainder of his clinical examination was unremarkable.

His complete blood count showed:

- Anemia with a hemoglobin level of 10.2 g/dL (normal range: 11.0-14.5 g/dL),

- Platelet count of 450 x 10^9/L (normal range: 150-450 x 10^9/L),

- White cell count of 2.8 x 10^9/L (normal range: 2.4-9.5 x 10^9/L).

The initial C-reactive protein level was elevated at 44. However, renal and liver function tests, bone profile, coagulation profile, and lactate dehydrogenase were all within normal limits. An admission chest X-ray revealed complete opacification of the right hemithorax with reduced lung volume and collapse. There was midline and tracheal shift to the left side, indicative of massive pleural effusion on the right side [Figure 1].

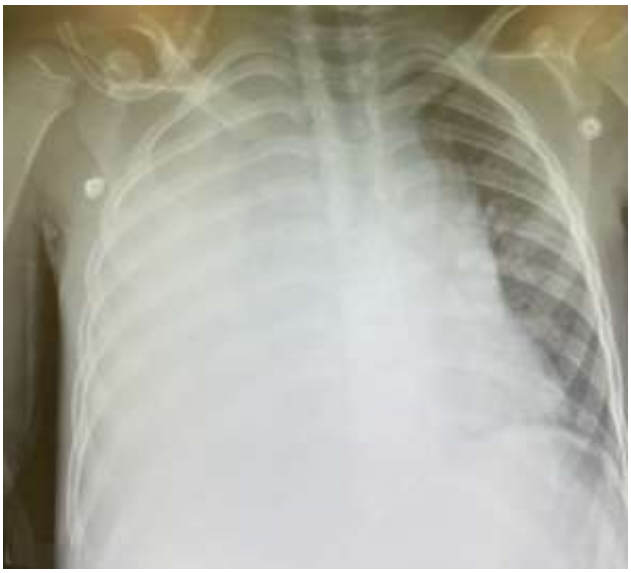

Figure 1: Chest X ray showing massive right side pleural effusion.

Point-of-care ultrasonography (POCUS) performed bedside revealed a right-sided pleural effusion. Initially admitted with a suspected diagnosis of massive pleural effusion possibly due to bacterial pneumonia, and received broad-spectrum antibiotics. An intercostal chest drain (ICD) was inserted, resulting in the drainage of a significant amount of fluid. [Figure 2].

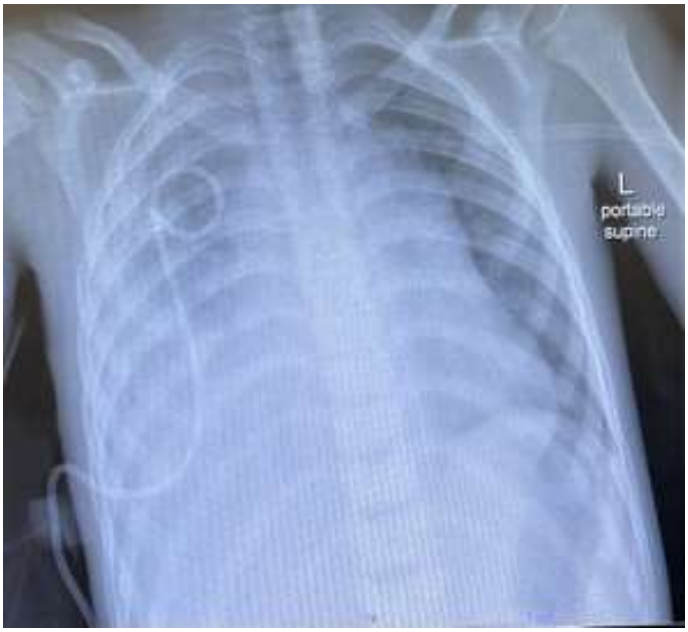

Figure 2: Chest X-ray after ICD insertion.

Following the insertion of the chest tube, the patient's clinical condition markedly improved. He was assessed by pediatric cardiologist and his echocardiography showed a small amount of pericardial effusion around the heart, with large anterior lobulated mass over the anterior mediastinum. Pleural fluid was sent for cytological examination, revealing monotonous infiltration with small to intermediate-sized cells. These cells exhibited a high nuclear to cytoplasmic ratio, irregular nuclear membrane, fine chromatin, and variable nucleoli. [Figure 3].

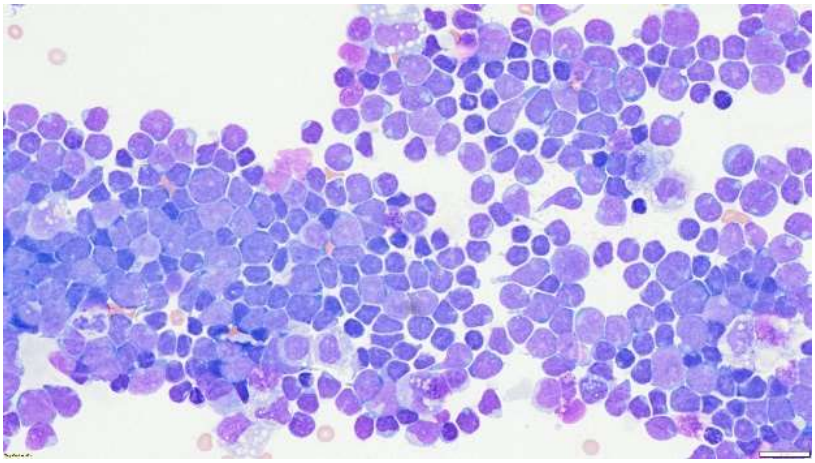

Figure 3: Pleural fluid cytospin showing small to intermediate-sized immature lymphocytes, with an open chromatin and scant cytoplasm. (May Grunwald Giemsa (MGG) Stain: 40x magnification).

Immunophenotyping by flowcytometry showed these to be T lymphoblasts (Positive for TdT, CD3, CD5, CD7, CD4, CD1a and CD99) as shown in figure 4. This was consistent with pleural involvement with T-cell lymphoblastic lymphoma/leukemia. Bone marrow aspirate and biopsy did not show any blast cells [Figure 4].

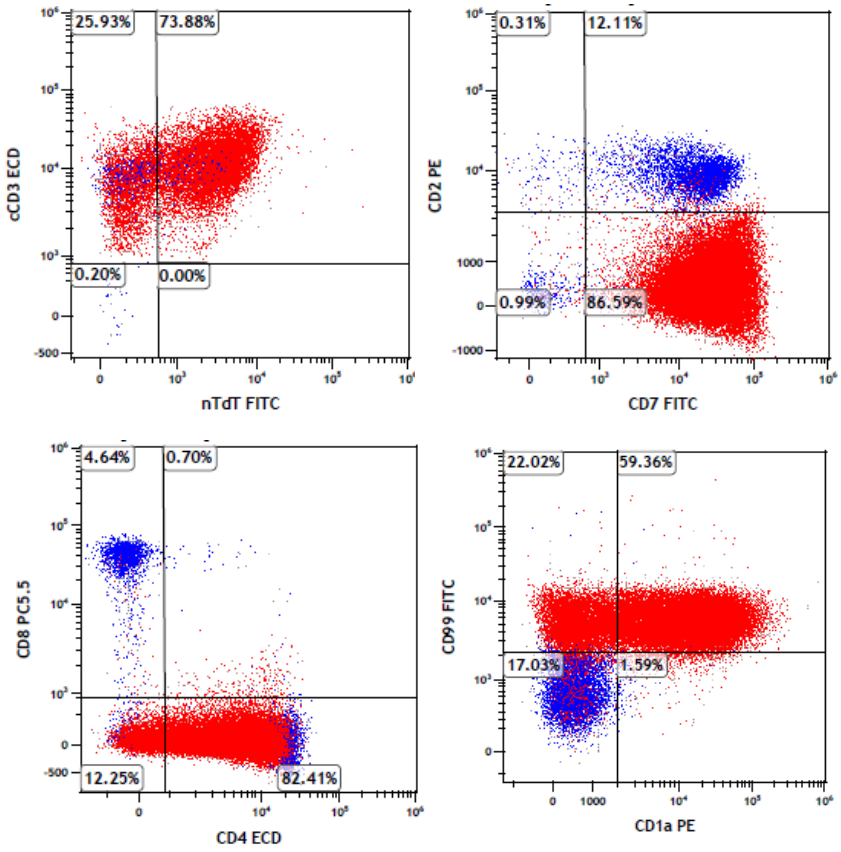

Figure 4: Immunophenotyping of pleural fluid showing T lymphoblasts (red events) which are expressing TdT, CD3, CD7, CD4, CD1a and CD99

Pan computed tomography scan revealed lymphomatous infiltrations of the thymus and hilar lymph nodes. A diagnosis of T-cell lymphoma was made based on the above findings, and chemotherapy treatment started based on COG AALL 1231.

Discussion

In our case, a 7-year-old previously healthy boy presented with acute onset of chest pain, lethargy, and shortness of breath, which prompted an investigation revealing massive pleural effusion and subsequent diagnosis of T-cell lymphoblastic lymphoma. This rare hematologic malignancy primarily affects children and adolescents, characterized by the aggressive proliferation of T-cell lymphoblasts in extramedullary sites such as lymph nodes, thymus, and occasionally, the pleural space.[1,2] The clinical presentation of our patient, including symptoms of respiratory distress and chest pain, is consistent with previous reports highlighting the potential for pleural involvement in T-cell lymphoblastic lymphoma/leukemia. Pleural effusion as an initial manifestation of hematologic malignancies, although uncommon, underscores the importance of considering malignancy in the differential diagnosis of pediatric patients presenting with atypical respiratory symptoms.[3]

Imaging studies played a crucial role in our patient's diagnosis and management. Chest X-ray initially demonstrated massive pleural effusion with mediastinal shift, providing the first indication of intrathoracic pathology.[4] Point-of-care ultrasonography (POCUS) confirmed the presence of pleural fluid and guided timely insertion of an intercostal chest drain, which facilitated symptom relief and sample collection for diagnostic cytology.[4] Computed tomography (CT) scan further delineated mediastinal lymphadenopathy and thymic involvement, allowing assessment of disease extent.[4] Although positron emission tomography (PET) scanning was not performed in this case, it is increasingly recognized for its role in precise staging, evaluation of treatment response, and detection of residual or

recurrent disease in T-cell lymphoblastic lymphoma.[4] This multimodal imaging approach, combined with cytological and immunophenotypic analysis, was essential for accurate diagnosis and guiding effective treatment.

While bone marrow involvement is common in lymphoblastic lymphoma/leukemia, extramedullary presentations without initial bone marrow infiltration, as observed in our patient's bone marrow aspirate and biopsy, highlight the importance of comprehensive diagnostic workup including imaging and cytology of affected sites.[5]

Treatment protocols for T-cell lymphoblastic lymphoma typically involve intensive, multi-agent chemotherapy regimens. In our case, the patient was started on the COG AALL1231 protocol shortly after diagnosis. Following induction, he showed marked clinical improvement with complete resolution of pleural effusion. At six months post-diagnosis, the patient remains in remission. The COG AALL1231 protocol has demonstrated favorable outcomes, with reported 5-year event-free survival rates of approximately 85% in pediatric patients with T-LBL.[4,6] This underscores the importance of early diagnosis and multidisciplinary management in optimizing prognosis for children with this aggressive malignancy.

In conclusion, our case illustrates the diagnostic challenges and therapeutic considerations in managing T-cell lymphoblastic lymphoma presenting with pleural effusion in a pediatric patient. Heightened clinical suspicion, integrated imaging techniques, and comprehensive cytological analysis are essential for timely diagnosis and initiation of appropriate treatment.

In a review of literature, similar cases of T-cell lymphoblastic lymphoma presenting with pleural effusion have been documented, emphasizing the variable clinical presentations and diagnostic complexities associated with extramedullary manifestations [Table 1]. Studies by Sandlund et al. have highlighted the importance of immunophenotyping in confirming T-cell lineage and guiding therapeutic strategies.[7]

Furthermore, reports by Liu et al. underscore the efficacy of intensive chemotherapy protocols like COG AALL 1231 in achieving remission and improving survival outcomes in pediatric patients with T-cell lymphoblastic lymphoma.[8]

These findings collectively support the approach taken in our case, reinforcing the significance of early recognition, accurate diagnostic techniques, and aggressive multimodal therapy in managing this rare and aggressive hematologic malignancy.

Table 1: Comparison of our case with previously reported pediatric T-LBL cases presenting with pleural effusion.

Feature	This Case (Current Report)	Case A (Liu et al., 2017)	Case B (Sandlund et al., 2009)
Age	7 years	9 years	10 years
Initial Presentation	Chest pain, SOB, massive pleural effusion	Pleural effusion and cough	Dyspnea, pleural effusion
Bone Marrow Involvement	None	Mild involvement	Present
Imaging	Chest X-ray, POCUS, CT	Chest X-ray, CT	Chest X-ray, PET-CT
Cytology Findings	Immature lymphoblasts	T lymphoblasts	T lymphoblasts
Immunophenotyping	TdT+, CD3+, CD7+, CD4+, CD1a+	TdT+, CD3+, CD5+	TdT+, CD3+, CD7+
Chemotherapy Protocol	COG AALL 1231	COG AALL 0434	BFM-based protocol
Outcome	Responded well, effusion resolved	Complete remission	Partial response

Conclusion

Malignancy should generally be considered a differential diagnosis in pediatric patients presenting with massive pleural effusion. If malignancy is suspected, fluid cytology should be conducted alongside basic workup and

radiological imaging. Additionally, bone marrow assessment and biopsy should be performed. While timely diagnosis can be challenging, it significantly improves the prognosis for these patients.

Disclosures

The authors declared no conflicts of interest. Written consent was obtained from the parents of the child to publish her photograph for research purposes.

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