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Acute Lymphoblastic Leukemia: A Case of Cardiac Tamponade as the Initial Presentation

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Abstract **Case Report**

Acute lymphoblastic leukemia (ALL) is a rare yet aggressive blood cancer predominantly affecting young adults, often presenting with symptoms related to cytopenias. While cardiac involvement in leukemia is uncommon, it can manifest as pericardial effusion (PE), occasionally leading to cardiac tamponade. We present a case of a 23-year-old male initially presenting with dyspnea and chest discomfort, later diagnosed with T-cell ALL following the discovery of PE. Urgent pericardiocentesis revealed leukemic blasts, confirming cardiac involvement. Despite prompt initiation of chemotherapy, the patient succumbed to complications, including febrile neutropenia and infectious pneumonitis. This case highlights the challenge of diagnosing ALL with atypical presentations and underscores the importance of a multidisciplinary approach for timely diagnosis and management, especially in high-risk patients where rapid deterioration can occur. Early recognition of cardiac involvement in leukemia is crucial for initiating appropriate treatment and improving outcomes.

Keywords: Acute Lymphoblastic Leukemia, Pericardial Effusion, Cytology.

INTRODUCTION

Acute lymphoblastic leukemia (ALL) is an uncommon yet aggressive blood cancer, mainly affecting young adults. It is believed to emerge from a complex combination of external or internal exposures, genetic factors, and random chance. The symptoms of (ALL) largely stem from cytopenias due to the buildup of leukemia cells in the body, manifesting as fever, fatigue, bone and joint pain, and a tendency to bleed[1, 2].

Commonly, leukemia can lead to complications such as pericardial effusion, which can vary in clinical presentation. Cases of leukemia presenting acutely with cardiac symptoms are exceedingly rare, with just a few instances documented in recent studies. This paper discusses a case where ALL initially manifested as cardiac tamponade.

CASE PRESENTATION

A 23-year-old male from a rural area was brought to the hospital presenting with dyspnea, cough, and escalating chest discomfort for 10 days, accompanied by night sweats and a general decline in health. The patient had no previous episodes of flu-like symptoms, fever, or exposure to tuberculosis, and no family members exhibited similar symptoms.

Upon examination, the patient was found to have acute orthopnea, with an oxygen saturation level of 88% on ambient air, blood pressure at 90/50 mmHg, a rapid heart rate of 150 beats per minute, and a paradoxical pulse. Signs indicative of right heart failure was present, such as evident jugular vein distension, oedema in the lower extremities, and notable ascites. Thoracic assessment showed dullness at the lung bases and subdued heart sounds without murmurs or friction rubs. Further examination revealed swollen, non-sensitive axillary lymph nodes, slight enlargement of the spleen, and petechiae on both legs, without any signs of skin or gingival infiltration.

Initial investigations, revealed abnormal blood counts: White Blood Cells (WBC) at 20G/L, Hemoglobin (HGB) at 10.1g/dL, and Platelets (PLT) at 23G/L. The blood smear performed reveals 6% NEUT, 4% LYMPH, and 90% of small to medium-sized lymphoblasts with high nucleocytoplasmic ratio, fine chromatin, and sometimes prominent nucleoli, all in favor of acute leukemia; bone marrow smears. The hemostasis workup was normal.

a chest radiograph, identified significant heart enlargement (Cardiothoracic Index [CTI] of 0.7) and moderate bilateral pleural effusions. Due to the patient's unstable hemodynamics, an urgent transthoracic echocardiogram (TTE) was conducted, confirming a substantial pericardial effusion. An emergency pericardiocentesis was promptly carried out, removing 1000ml of hemorrhagic fluid, which upon analysis showed an exudative nature with the presence of leukemic blasts (Figure 1). Post-procedure, the patient showed clinical improvement, corroborated by follow-up echocardiography.

Bone marrow analysis and immunophenotyping confirmed the diagnosis of T-acute lymphoblastic leukemia (Figure 2). and immunophenotyping revealed an aspect of T-acute lymphoblastic leukemia, and the chromosomal formula on the hematological karyotype (according to ISCN 2016) was 47, XY, +3 del [9], (q22q32), -11, der (14) t (14) (q32), +mar (7)/46, XY (27); The FISH was pending.

The patient commenced treatment with prednisone (2mg/kg/day) and allopurinol (400mg daily) as initial therapy. Further investigations for infectious diseases (including tuberculosis and COVID-19), autoimmune disorders, and solid cancer markers, alongside comprehensive liver, kidney, and thyroid function tests, returned negative results. After undergoing a cycle of cytoreductive chemotherapy (COP), the patient showed some hematological improvement. However, persistent symptoms such as lymphadenopathy and pleural effusion led to the decision to start the GRAALL2013 treatment protocol, even before the FISH results were available. Unfortunately, on day five of the chemotherapy regimen, the patient developed febrile neutropenia accompanied by respiratory distress and widespread petechial purpura. These complications rapidly worsened, culminating in the patient's demise despite intensive supportive care measures. Further investigations during the treatment phase revealed severe infectious pneumonitis amid acute renal failure as complicating factors.

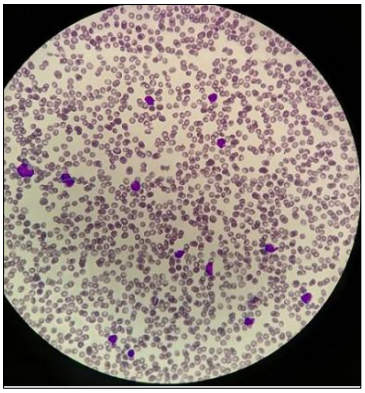

Figure 1: blasts in the pericardial fluid

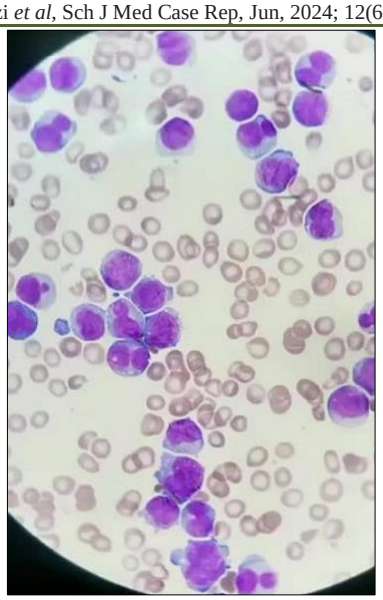

Figure 2: bone marrow aspirate smear: the marrow is invaded by lymphoblasts of various sizes, with regular nuclei and fine chromatin No myeloid or erythroid precursors are seen.

DISCUSSION

The occurrence of PE in leukemia patients is increasingly recognized, Although poorly understood, the pathophysiology behind the relationship between leukemia and acute pericardial disease is thought to be related to multiple factors including hemorrhage caused by concurrent thrombocytopenia, infections due to underlying immune deficiency, and direct malignant cell infiltration [3, 4]. While the prognostic and therapeutic implications of malignant effusions in ALL are not entirely known, it appears that an effusion large enough to cause tamponade increases mortality [5].

Although PE as an initial symptom of leukemia has historically been rare, recent studies suggest that a substantial proportion of symptomatic effusions without a clear cause may be the first indicator of undiagnosed cancer, with malignancies more commonly being metastatic rather than primary cardiac tumors [6]. Despite the unusual presentation, our patient's diagnosis of acute lymphoblastic leukemia was swiftly established, after ruling out other potential differential diagnoses, particularly tuberculosis. The T-cell type of ALL is a malignant disorder characterized by the massive clonal proliferation of immature lymphoid progenitor cells, as per the 2016 WHO classification of lymphoblastic leukemia/lymphoma [7]. The diagnosis is primarily based on the demonstration of surface/cytoplasmic CD3, as most T-cell markers are not entirely lineage-specific. Cytogenetics and analysis of cellular DNA content are utilized to identify submicroscopic leukemia type-specific abnormalities and guide therapy [1]. All structural abnormalities observed in our patient´s karyotype (trisomy 3 and del 9q) are reported in type T ALL.

Effusion cytology aims to achieve an early and quick diagnosis of malignancy. In our patient, cytological studies of the pericardial fluid were positive. The diagnosis in such cases requires correlation of various cytomorphologic features, immunocytochemistry (ICC) and patient's clinical details [8]. In a retrospective study, 51 positive cytological fluid samples from patients with lymphoreticular malignancies that included 30 (58.8%) pleural, 18 (35.3%) peritoneal, and 3 (5.9%) pericardial, were also analyzed by flow cytometry (FCM) and/or ICC10. Forty-eight samples were diagnostically confirmed by using theses techniques, but 3 (5.9%) were considered to be false positives since FCM and/or ICC did not support a lymphoreticular malignancy [9].

Immediate, tailored treatment initiation is critical. Pre-phase therapy aims to reduce tumor burden and prevent tumor lysis syndrome, followed by induction therapy to achieve complete remission with minimal toxicity. The selection of the GRAALL protocol reflects evidence suggesting better outcomes with pediatric-based regimens for adolescents and adults with ALL [10].

ALL's pathological process leads to bone marrow failure and severe cytopenia, with untreated cases being rapidly fatal. Multi-drug chemotherapy introduces the risk of myelosuppression and complications such as tumor lysis syndrome and febrile neutropenia, necessitating vigilant management and prompt intervention for infections [11]. The presented case underscores the challenges of managing a rapidly progressing disease resistant to initial therapy and complicated by severe infectious pneumonitis, contributing to a poor prognosis.

CONCLUSION

This case report illustrates a rare and early manifestation of acute lymphoblastic leukemia (ALL) in a patient initially presenting with cardiac tamponade, without prior indications of cancer. It underscores the importance of considering ALL in the differential diagnosis for new-onset pericardial effusion.

The case further demonstrates the value of an interdisciplinary approach, combining expertise from various specialties for swift diagnosis, effective chemotherapy delivery, and comprehensive supportive care. This approach is especially vital for managing high-risk patients, where conditions can deteriorate quickly, emphasizing the need for attentive and all-encompassing medical care.

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