A CASE WITH ACUTE LEUKEMIA PRESENTING WITH CARDIAC TAMPONADE*

*Ayhan Söğüt MD**, Kutluhan Yılmaz MD**, Nevin Yalman MD****

*Kamil Şahin MD***, Mustafa Babalıoğlu MD*****

*Rukiye Eker Ömeroğlu MD*****, Talat Cantez MD******

SUMMARY: Söğüt A, Yılmaz K, Yalman N, Şahin K, Babalıoğlu M, Ömeroğlu RE, Cantez T. (Department of Pediatrics, İstanbul University Faculty of Medicine, Çapa-İstanbul, Turkey). A case with acute leukemia presenting with cardiac tamponade. Turk J Pediatr 1999; 41: 509-516.

Pericardial effcisions and cardiac tamponade are rare and severe complications of leukemia. They often develop during the radiation therapy, chemotherapy, or infections in the course of leukemia. However, some cases present with pericardial effusion and tamponade. We report a three-year-old girl who was admitted with cardiac tamponade and needed urgent pericardiocentesis. Clinical evaluation and laboratory results revealed myeloid markered-T cell acute lymphoblastic leukemia (ALL) and pericardial invasion. She is the youngest patient with cardiac tamponade who was diagnosed acute lymphoblastic leukemia in the English-language literature. *Key words: pericardial effusion, acute lymphoblastic leukemia.*

A variety of cardiac and noncardiac conditions such as congestive heart failure, infections, autoimmune and renal diseases and cardiac surgery may cause pericardial effusion. Leukemia is one of the rare causes of pericardial effusion. It occurs mainly as a consequence of chemotherapy, radiotherapy, or of infections while treating patients with leukemia. However, pericardial effusion is rarely the initial presentation of acute leukemia. There are 12 cases who presented with cardiac tamponade as the initial manifestation of acute leukemia in English-language literature[4-14]. We present a three-year-old girl who was referred because of cardiac tamponade. She needed urgent pericardiocentesis and then an acute lymphoblastic leukemia (ALL) and pericardial invasion were determined.

Case Report

A three-year-old girl whose complaints were cough, fever, dyspnea and swelling of the eyelids was refferred to our hospital prediagnosed as pericarditis.

* From the Department of Pediatrics, İstanbul University İstanbul Faculty of Medicine, Çapa-İstanbul.

** Research Assistant in Pediatrics, İstanbul University İstanbul Faculty of Medicine.

*** Associate Professor of Pediatrics, İstanbul University Istanbul Faculty of Medicine.

**** Pediatrician, Leukemia Foundation Health Unit, İstanbul

***** Professor of Pediatrics, İstanbul University Istanbul Faculty of Medicine.

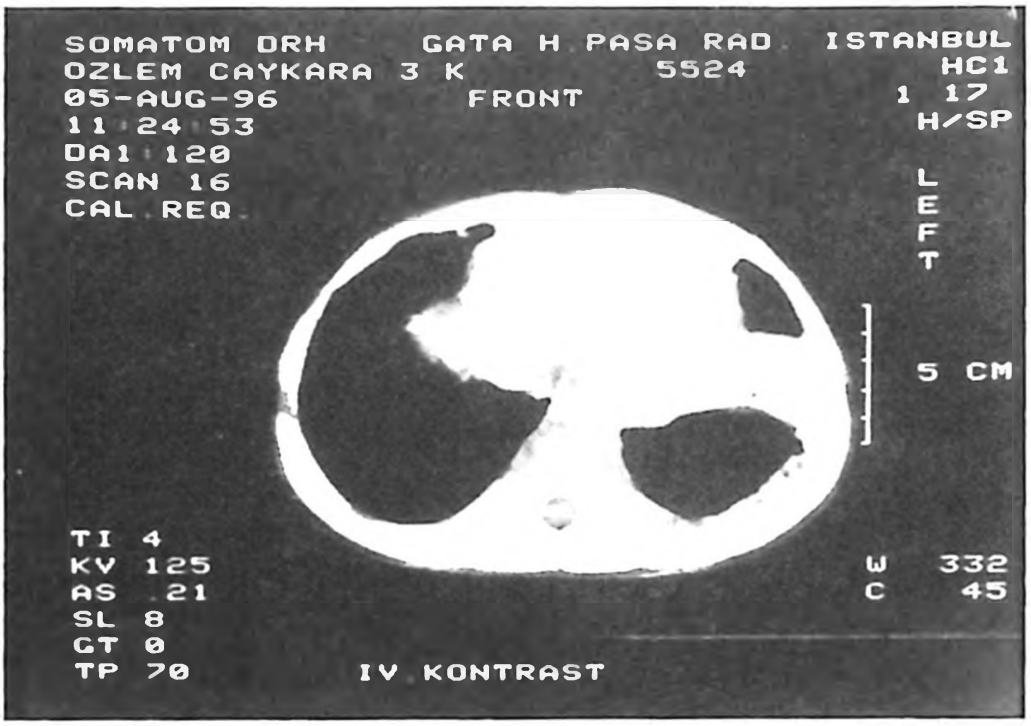

Fig. 1: Pericardial and pleural effusion in chest tomography.

The patient had been treated with iron tablets because of pica and iron deficiency for a year. She was brought to a local hospital one month previously with complaints of cough, fever, dyspnea and fatigue. Ceftriaxone was administered for the diagnosis of bacterial bronchopneumonia. However, an enlargement of the cardiac silhouette was noticed in control chest X-rays. Echocardiography, thoracal and abdominal ultrasonography and chest tomography (Fig. 1) revealed pericardial and minimal pleural effusion and mild hepatosplenomegaly, but no mediastinal or abdominal lymphadenopathy or mass. Other laboratory findings were as follows: hemoglobin: 8.1 g/dl; white blood cell count: 8000/mm^3 with 60 percent lymphocytes, 4 percent reactive lymphocytes (not blasts), 30 percent neutrophils, 4 percent band forms, 2 percent others; platelet count: 112,000/mm^3 and erythrocyte sedimentation rate: 65 mm/h. She had been treated at the local hospital for bronchopneumonia for 10 days without success and, when she deteriorated, was sent to our hospital for further evaluation.

Upon admission to our hospital, pallor, restlessnes, tachypnea, dyspnea, sweating and periorbital edema were noticed on physical examination. Her pulse rate was with 160/min and blood pressure was 90/60 mmHg, and she had marked jugular vein distension. Heart sounds were weak, but there were no murmurs or friction rubs. Crackling rales were heard over basal areas of both lungs. The liver was 4 cm and the spleen 3 cm palpable below the respective costal margins. The rest of the physical examination was normal.

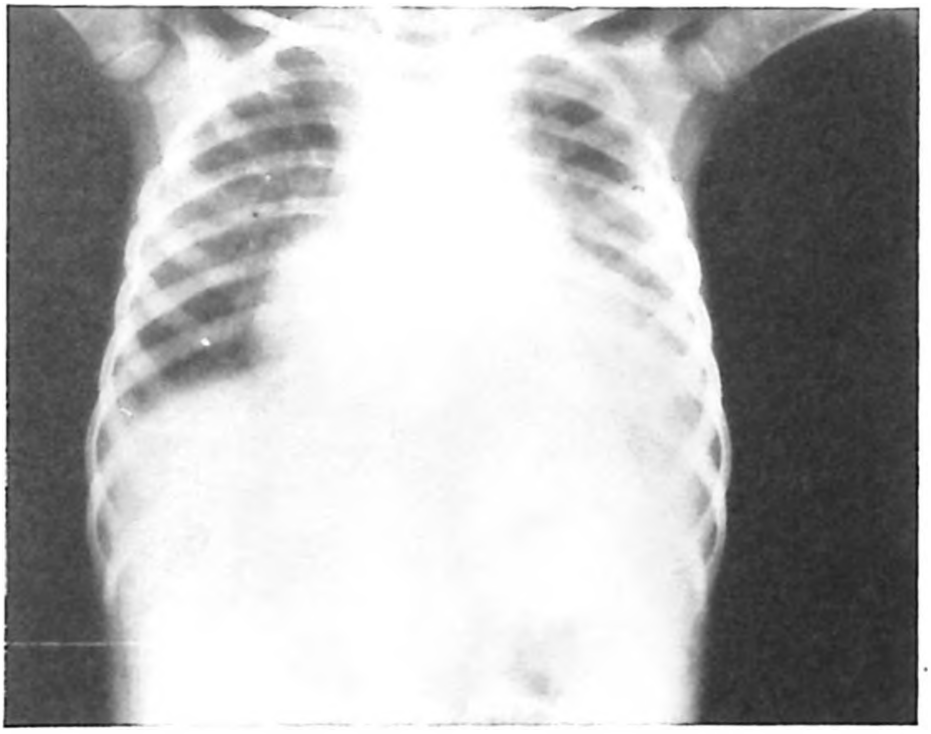

Fig. 2. Cardiomegaly in telecardiography before pericardiocentesis.

The telecardiography before pericardiocentesis showed a marked cardiac enlargement (Fig. 2). Sinus tachycardia, low QRS voltages and negative T waves on V_{1-6} were present in ECG tracings. On echocardiography, pericardial effusion, collapse of right atrial wall and anterior wall of right ventricle during diastole period was present (Fig. 3). Minimal pleural effusion, bilateral but more apparent

Fig. 3. Pericardial effusion on echocardiography

on left side, was also observed by thorax ultrasonography. Clinical and echocardiographical evaluation revealed a pericardial tamponade requiring an urgent pericardiocentesis. Pericardiocentesis yielded 200 ml of serohemorrhagic fluid. Fluid analysis revealed glucose 74 mg/dl, protein 2.85 g/dl, LDH 4,000 IU/L, simultaneous serum protein 6.3 g/dl and LDH level 2,872 IU/L. No

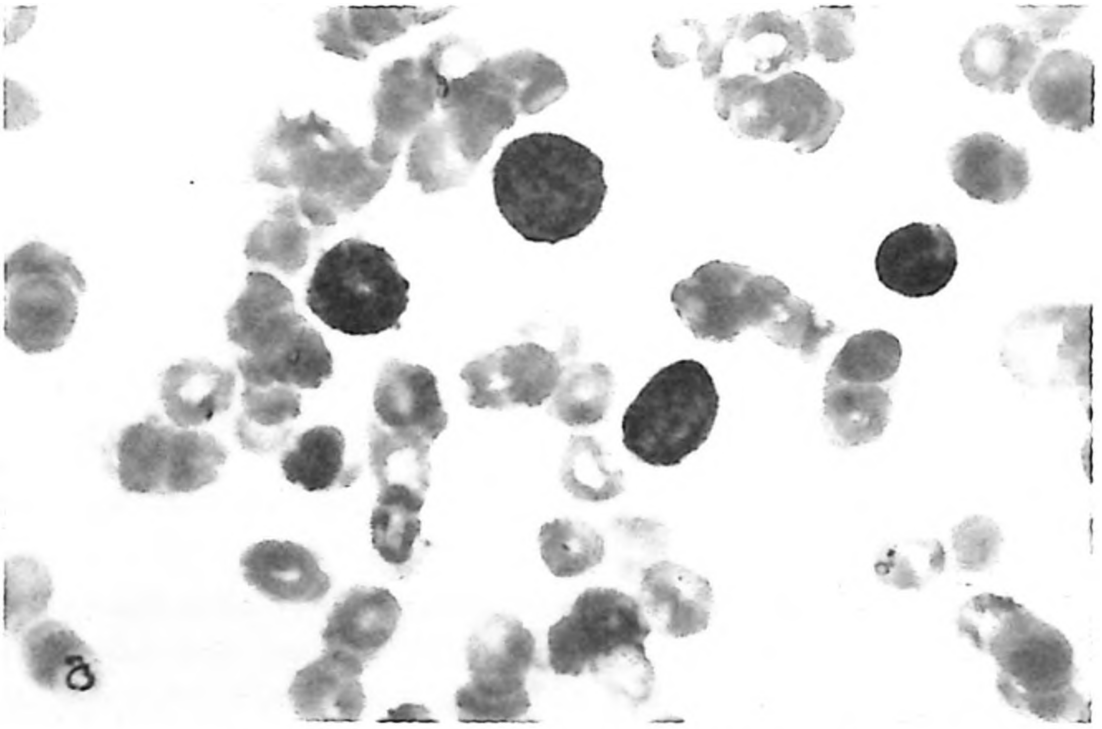

Fig. 4: Lymphoblastic cells in pericardial fluid.

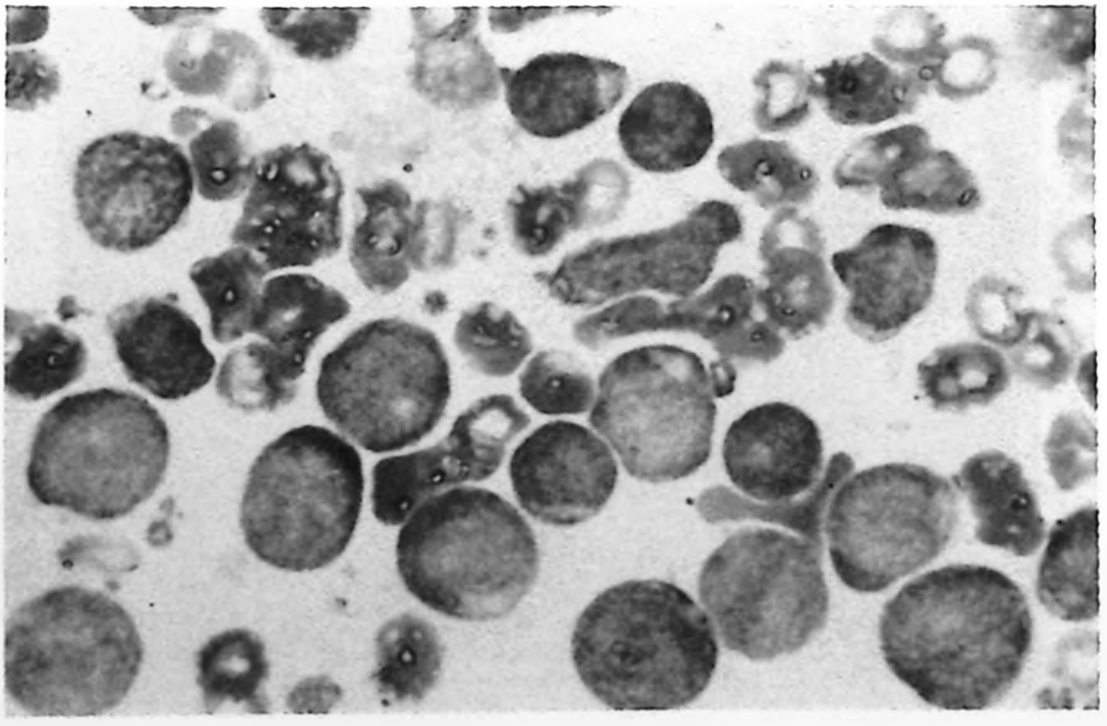

Fig. 5: L_2 type lymphoblasts on bone marrow.

microorganisms or M. tuberculosis were detected in the fluid. Among cells, 25 percent microlymhoblastoid cells were observed in the pericardial fluid (Fig. 4). Hematological values were as follows: hemoglobin 6.3 g/dl; hematocrit 19.7 percent; mean corpuscular volume (MCV) 81 fl; platelet count 180,000/mm^3; and white blood cell count 16,000/mm^3 with 20 percent neutrophils, 70 percent lymphocytes, and 10 percent lymphoblasts. Blood urea, ALT, AST and serum electrolytes and routine urine examination were all normal. Bone marrow aspiration revealed a hyperplastic marrow with L_2 type mononuclear cell infiltration (Fig. 5). CALLA, CD 19, CD 20, CD 4, CD 8, CD 13, CD 14 were negative and CD 2, CD 7, CD 34, CD 33 were positive by immunohistochemical and immunophenotypical studies in bone marrow and, following these results, myeloid markered-T cell ALL was diagnosed. Central nervous system involvement was not detected. She was administered the augmented BFM-86 CCG-1882 chemotherapy protocol. Pericardial and pleural effusion disappeared two weeks later (Fig. 6) and, after one year, her bone marrow is still in remission.

Discussion

Cardiac involvement is a well-known feature in hematological malignancies, and it has been reported at autopsy in approximately 20 percent of patients with lymphomas or leukemias[1]. Most are asymptomatic. Infiltration of the pericardium is a much more common finding in postmortem examinations of patients with acute leukemia (AL)[2]. Generally, pericardial involvements detected before death

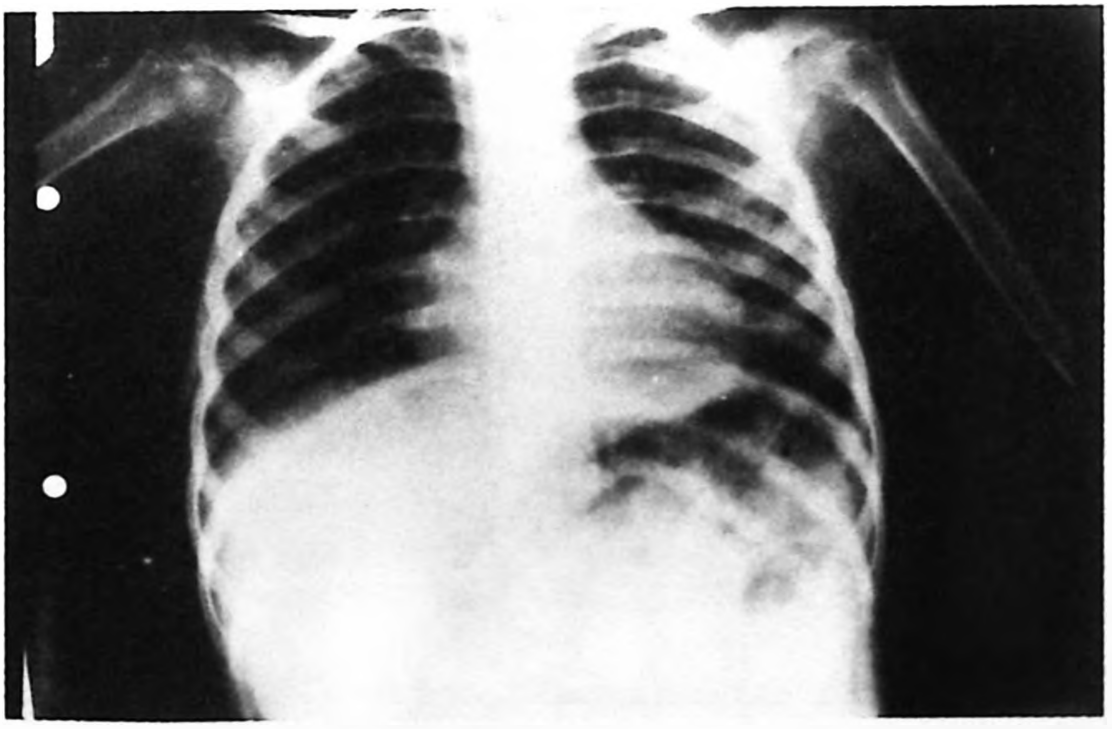

Fig. 6. Normal telecardiography after chemotherapy

have been diagnosed in the advanced stages of neoplastic blood diseases[2,3]. In contrast to the high frequency of pericardial involvement in the course of AL, pericardial effusion is rarely present at the onset of AL, and is rarely the initial manifestation of AL. The occurrence of pericarditits after bronchopneumonia without any finding suggesting acute leukemia was also noted in our case. Although our case developed cardiac tamponade, she did not demnostrate any abnormality in hematological parameters or imaging consistent with AL in the beginning. The patient was admitted to our hospital with severe cardiac tamponade. An emergency pericardiocentesis was performed and microlymphoblastoid cells were detected on the smear of the pericardial effusions. Thereupon, a peripheral blood smear and bone marrow aspiration were done, and blastic cells were seen on both.

Battle et al.[13] reported a review in 1991 containing 10 patient reports plus their own patient, and Spottswood et al.[14] also reported another case. The age range was two to 45 years. Four cases had acute myeloblastic leukemia and nine acute lymphoblastic leukemia; our case is the youngest patient with acute lymphoblastic lekemia. A summary of these cases is present in Table I.

Table I: ALL Cases with Pericardial Involvement in English-Language Literature

	No of Reference	Age (year)	Sex	Type of AL	Blasts in the Pericardial Effusion
Rab et al.[4]	4	26	M	ALL	Present
Battle et al.	5	6	F	ALL	Present
Jaffe et al.	6	15	F	ALL	Present
Jaffe et al.	6	5	M	ALL	Not Performed
Chia et al.	7	19	F	ALL	Present
Krause	8	16	M	AML	Present
Sobolewski et al.	9	12	M	ALL	Not Perfomed
Chu et al.	10	5	M	AML	Present
Mancuso et al.	11	18	F	ALL	Absent
Leung et al.	12	45	F	ALL	Not Performed
Battle M et al.	13	18	F	AML	Present
Spottswood et al.	14	2	F	AML	Present
Our case et al.		3	F	ALL	Present

Abbreviations: AL: acute leukemia; ALL: Acute lymphoblastic leukemia; AML: Acute myeloblastic leukemia; F: Female; M: Male.

Pericardiocentesis was performed on nine patients. Blastic cells were detected in eight of nine pericardial fluid samples. Pericardial effusion responded to chemotherapy and did not recur, although AL relapsed. We had to perform pericardiocentesis because of our patient's cardiac tamponade. We diagnosed the same day and started to administer chemotherapy four days later. Pericardial effusion resolved in two weeks and did not reaccumulate.

The optimal management in cardiac tamponade as a result of leukemic infiltration is still controversial. Pericardiocentesis is only to be performed to relieve the patient with cardiac tamponade and to attain hemodynamic stabilization. Systemic chemotherapy is the essential therapeutic approach. Although surgical drainage with subxiphoid pericardiotomy might be recommended to prevent the reaccumulation of fluid or to evacuate the reaccumulated fluid, perhaps the best approach is drainage followed by rapidly systemic chemotherapy. The prognosis depends on the leukemia itself, rather than on pericardial involvement.

The cases mentioned above demonstrate that, although rare, leukemia my present with cardiac tamponade. Careful cytologic examination of pericardial fluid peroxides valuable diagnotic information in such cases.

REFERENCES

1. Terry LN Jr, Kligerman MM. Pericardial and myocardial involvement by lymphomas and leukemias. Cancer 1970; 25: 1003-1008.
2. Roberts WC, Bodey GP, Wertlake PT. The heart in acute leukemia. A study of 420 autopsy cases. Am J Cardiol 1968; 21: 388-412.
3. Sumners JE, Johson WW, Ainger LE. Childhood leukemic heart disease. A study of 116 hearts of children dying of leukemia. Circulation 1969; 40: 575-581.
4. Rab SM, Yee A. Initial cardiac tamponade in acute leukemia. Br Med J 1967; 1: 612.
5. Battle CU, Bonfiglio TA, Miller DR. Pericarditis as the initial manifestation of acute leukemia: report of a case. J Pediatr 1969; 75: 692-694.
6. Jaffe N, Traggis DG, Tefft M. Acute leukemia presenting with pericardial tamponade. Pediatrics 1970; 45: 461-465.
7. Chia BL, Da Costa JL, Ransome GA. Cardiac tamponade due to leukemic pericardial effusion Thorax 1973; 28: 657-659.
8. Krause JR. Granulocytic sarcoma preceding acute leukemia. A report of six cases. Cancer 1979; 44: 1017-1021.
9. Sobolewski S, Sreharan N, Rajah SM. Successful management of cardiac tamponade in two cases of leukemia. Br Med J 1979; 2: 1190.
10. Chu JY, Demello D, O'Connor DM, Chen SC, Gale GB. Pericarditis as presenting manifestation of acute nonlymphocytic leukemia in a young child. Cancer 1983; 52: 322-324.
11. Mancuso L, Marchi S, Giuliano P, Pitrolo F. Cardiac tamponade as first manifestation of acute lymphoblastic leukemia in a patient with echcgraphic evidence of mediastinal lymph nodal enlargement. Am Heart J 1985; 110: 1303-1304.

12. Leung WM, Tai YT, Lau ChP, Wang ChK, Cheng ChH, Chan TK. Cardiac tamponade complicating leukemia: immediate chemotherapy or pericardiocentesis? Postgr Med J 1989; 65: 773-775.

13. Battle M, Ribera JM, Larrousse E, et al. Cardiac tamponade as the initial manifestation of acute leukemia: report of a case and review of the literature. Haemotologica 1991; 76: 505-507.

14. Spottswood SE, Goble MM, Massey GV, Ben-Ezra JM. Acute monoblastic leukemia presenting with pericardial effusion and cardiac tamponade. Pediatr Radiol 1994; 24: 494-495.