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Case Reports

Pericardial Tamponade in Acute Myeloid Leukemia

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We are reporting a case of acute myeloid leukemia in a child with pericardial effusion and cardiac tamponade, with a review of the literature. Morphologic and cytochemical evidence was used to diagnose acute myeloid leukemia, and radiologic, electrocardiographic, echocardiographic, cytologic, and histologic evidence confirmed the diagnosis of tamponade and pericardial leukemic involvement. Pericardiotomy and pericardial window drainage successfully relieved the tamponade, and conventional chemotherapy was used to induce a remission.

In the terminal stages of leukemia, leukemic involvement of the heart and pericardium is common, but as an initial manifestation of leukemia, involvement of the pericardium is rare in adults and almost unknown in children. In a study of 491 cases of childhood leukemia in Denmark (1), no pericarditis was present at the time of diagnosis.

This report concerns the case of a 14-year-old girl who had signs and symptoms of pericardial effusion when acute myeloid leukemia (AML) was diagnosed. She developed cardiac tamponade, which required pericardiectomy and drainage in association with chemotherapy and radiotherapy.

Case Report

A 14-year-old white girl presented to the Emergency Room in July 1979 with a history of epistaxis, sore throat, abdominal pain, vomiting of "coffee ground" material, fatigue, tendency to bruise easily, and fever of one week's duration. Two weeks before she was admitted, the mother noted bruising, decreased appetite, and lethargy. A dose of penicillin was administered at an outside clinic three days before admission. Her past history revealed two hospitalizations for joint pain in 1973, at which time a diagnosis of rheumatic fever with carditis had been made. At that time, prophylactic penicillin was begun. Her cardiac status remained normal without symptoms or any evidence of cardiac murmurs. Her family had a significant history of hypertension, as well as renal disease in a brother and von Willebrand's disease in a niece.

Physical examination revealed a pale, lethargic adolescent with a temperature of 35.8°F, pulse 120/minute, respiratory rate 40/minute, with moderate respiratory distress, orthopnea, and a blood pressure of 120/60. Extensive ecchymoses were noted on the legs, arms, and chest. Marked anterior and posterior cervical adenopathy was present bilaterally. Ophthalmoscopy revealed bilateral retinal hemorrhages. Jugular venous pressure was 12 cm without Kussmaul's sign. Carotid pulsations were prominent, and all peripheral pulses were palpable. When her blood pressure was measured, a paradox of 14 mmHg was evident. S_1 was normal, S_2 was split and mobile, and no S_3 or S_4 was heard. A Grade II/VI ejection systolic murmur was heard best along the left parasternal second and third spaces. No pericardial rub was aubible. Decreased breath sounds were noted in the left interscapular area paraspinally (Ewart sign). The liver, which was palpable 7 cm below the costal margin, was smooth, soft and nontender. No hepatojugular reflux could be demonstrated. The spleen was palpable 4 cm below the costal margin. Neurological examination was within normal limits.

Laboratory investigations on admission revealed a hemoglobin of 6.4 gm/100 ml, hematocrit of 17%, platelet count of 9,000/cu mm, and white cell count of

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170,000/cu mm. Differential count revealed 50% myeloblasts (many with Auer rods), 10% polymorphonuclear leukocytes, 10% bands, and 20% lymphocytes. Urinalysis and electrolytes were normal. BUN was 26 mg/dl (normal, 10-26); creatinine, 1.4 mg/dl (normal, 0.6-1.3); PT, 13.5 seconds (control, 12 seconds); PTT, 30 seconds (control, 26-42 seconds); LDH, 1201 u/l (normal, 166-290); SGOT, 165 u/l (normal, 9-33); SGPT, 97 u/l (normal, 4-40); uric acid, 10.1 mg/dl (normal, 2.5-8); calcium, 7.7 mg/dl (normal, 8.5-10.5); and phosphate, 5.6 mg/dl (normal, 2.5-4). Blood glucose, total protein, albumin, globulin, and triglycerides were within normal limits. Chest radiograph revealed cardiomegaly with a small left lower lobe infiltrate (Fig. 1). Bone marrow examination revealed 100% cellularity with 54.6% granulocytic stem cells, Auer rods, Reider myeloblasts, and micromyeloblasts. Cytochemical analysis indicated peroxidase specific esterase and Sudan Black positivity. Nonspecific esterase was positive in 5% of the cells, and the periodic acid Schiff test was negative.

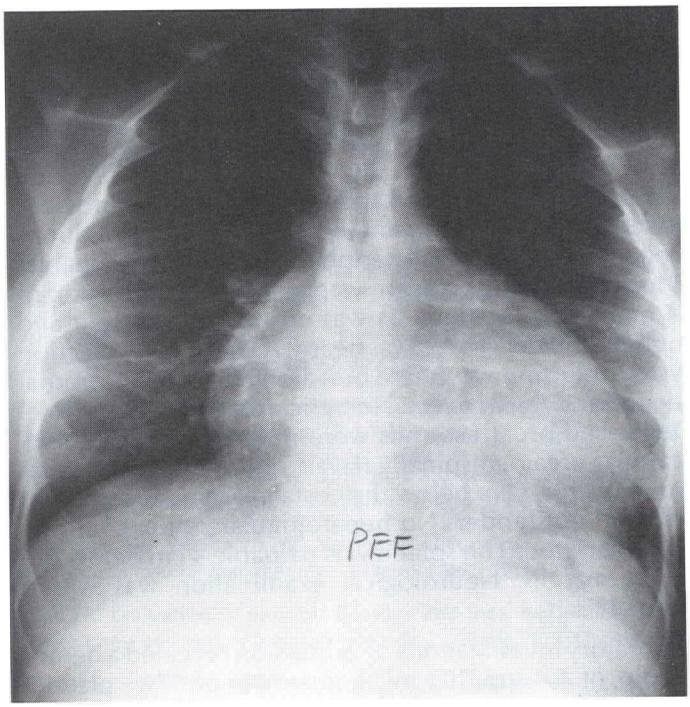

Fig. 1
Chest radiograph indicating enlarged heart.

Based on the bone marrow examination, the diagnosis was acute myeloid leukemia, M_2FAB (2). Chromosomal analysis of the bone marrow revealed translocation between chromosome 8 and 21. The karyotype analysis of 13 metaphase cells indicated that all cells were abnormal; 46XX, t(8;21) (q22;q22) was found in seven or more cells.

The patient was admitted to the pediatric intensive care unit and treated with vincristine, hydrocortisone, allopurinol, packed red blood cells, and platelets. Ampicillin and gentamycin were administered intravenously after appropriate cultures had been taken. The electrocardiogram showed decreased voltage, elevated ST segment, and electrical alternans. The paradoxical pulse increased to 22 mmHg, and the patient continued to be tachypneic and tachycardic. Urine output decreased, and blood urea nitrogen increased to 37 mg/dl, creatinine to 1.6 mg/dl. A repeat chest radiograph revealed increased cardiac enlargement and pulmonary congestion. Echocardiogram revealed significant anterior and posterior pericardial effusion (Fig. 2). A Swan-Ganz catheter introduced for monitoring revealed the following values: right atrium pressure, 16 mmHg; pulmonary wedge pressure, 19 mmHg; cardiac index, 2.11/min/m^2. When the patient's blood pressure began to fall and her urinary output began to drop, she was taken to the operating room.

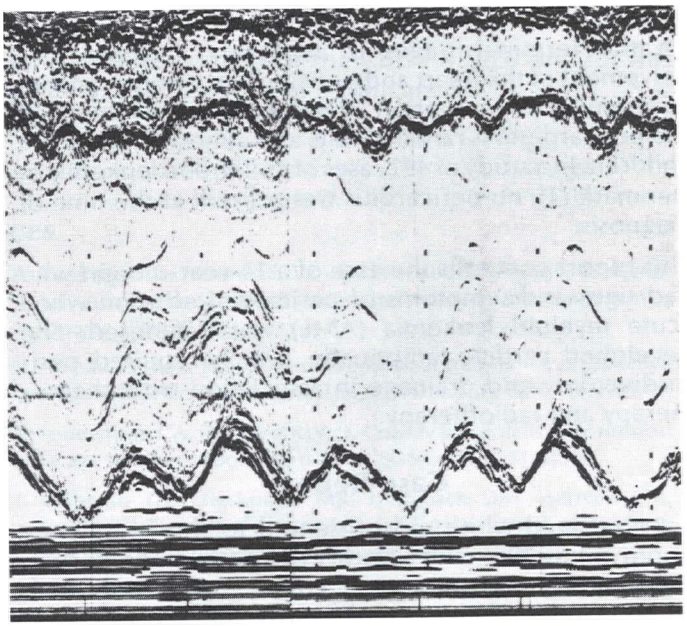

Fig. 2
Echogram of the heart indicating evidence of anterior and posterior pericardial effusion.

Through a left anterior thoracotomy, a pleuropericardial window was created. The pericardium was found to be tense, and when it was incised, the pericardial fluid was clearly under high pressure. Pericardial biopsy was obtained, and 500 cc of serosanguinous fluid was drained. The fluid showed a hematocrit of 15%, white blood cell count of 101,500 cu/mm with a differential count of 8% polycytes, 4% bands, 28% metamyelocytes, 9% monocytes, 28% myeloblasts, 7% myelocytes, and 14% lympho-

cytes. Bacterial, AFB, fungal, and viral cultures of the pericardial fluid were negative. The pericardial biopsy showed a leukemic infiltrate. When the pericardial pressure was relieved, the patient's systemic arterial pressure rose, and her pulmonary arterial pressure fell.

Postoperatively, the size of the cardiac silhouette decreased, tachypnea and orthopnea were relieved, and the urine output increased. The patient received conventional doses of vincristine, cytosine arabinoside, and prednisone for induction chemotherapy. Hematological remission was achieved without difficulty.

Three months after her remission began, reaccumulation of pericardial fluid without cardiac tamponade was detected by radiographic and echographic tests, but pericardiocentesis was not attempted. Her bone marrow was in remission at that time, and over 22 days she received 2,100 rads to her heart by means of a 6 MEV x-ray machine and parallel opposing anterior and posterior beams. Her remission continued with vincristine, prednisone, 6-MP and methotrexate therapy.

Fifteen months after initial diagnosis, a leukemic infiltrate was found in the right breast and confirmed by biopsy. At this time, the bone marrow showed 7% myeloblasts. The patient was again treated with reinduction chemotherapy using vincristine, cytosine arabinoside, and prednisone; she also received 2,600 rads to the right breast. The breast mass completely disappeared after irradiation and chemotherapy.

However, the bone marrow relapsed 27 months later, and the patient died in November 1981. At this relapse, chromosomal analysis of bone marrow revealed five of 12 G banded metaphase cells with karyotype of 46XX; t(8;21) (q 22; q 13). The other seven had 47XX + 1 (t 8;21) (q 22; q 13). Autopsy demonstrated extensive leukemic infiltration of meninges, heart, lung, liver, spleen, lymph nodes, and bone marrow. Petechial hemorrhages were present over the entire body, including the brain. Postmortem blood cultures were positive for candida albicans.

Discussion

Cardiac tamponade is an unusual manifestation of acute myeloid leukemia (3). In the study by Koch, et al (4) of 192 cases of acute leukemia in children, no child had pericarditis as a complication. Battle, et al (5) and Jaffe, et al (6) reported one child and two children, respectively, with acute lymphocytic leukemia and pericardial effusion at the time of initial diagnosis. Before these reports, only five cases had been reported, the youngest patient being 15 years old (7-12). Rab and Yee (13) described a 26-year-old man with acute lymphocytic leukemia and cardiac tamponade at the time of diagnosis who was treated with corticosteroids, 6-mercaptopurine, and repeated pericardiocentesis. Chia, et al (14) treated a 19-year-old woman for tuberculosis because the initial evaluation of both pericardial fluid and peripheral blood failed to reveal leukemia. However, when cardiac tamponade recurred after three months, the pericardial fluid and bone marrow revealed lymphoblasts.

Pipoly and Rogers (15) discussed the presentation and management of a 14-year-old child with an unusual myeloproliferative disorder characterized by hepatosplenomegaly, myelofibrosis, ringed sideroblasts, and conversion of acute myeloblastic leukemia. The patient developed cardiac tamponade secondary to extramedullary hematopoiesis.

The diagnosis of AML in our patient was confirmed by morphological and cytochemical studies of peripheral blood and bone marrow. The chromosomal analysis with G banding studies indicated an 8 to 21 translocation which has been called the "prototype" karyotype and may even be a "classical" karyotype for AML (16). It has also been reported that the presence of this karyotype may be correlated with the patient's long survival (16,17). The cell line with trisomy 1 was seen only at the last relapse and represented the evolution of the leukemia. Trisomy 1, which occurs in many solid tumors, is seen only infrequently in leukemia and is an unfavorable prognostic sign.

The presence of cardiac tamponade was suggested by the clinical findings of distant heart sounds, cardiomegaly, Ewart's sign, pulsus paradoxus, and the absence of S_3 despite overt left heart failure. The diagnostic echocardiogram was important in the management of the tamponade and in detecting the degree of effusion. The Swan-Ganz catheter and hemodynamic measurements performed in this child may not always be necessary to confirm the diagnosis. Before this invasive procedure is performed, the risks of hemorrhage due to low platelets and of secondary infection in an immunocompromised and neutropenic host must be carefully considered. Aggressive, invasive monitoring and surgical intervention with pericardiotomy and drainage through a pericardial window were well tolerated by the patient. Successful emergency management of pericardial effusion employed intravenous hydrocortisone and mediastinal irradiation (total doses ranging from 500 to 1,160 rads) along with conventional antileukemic therapy (3,4,6). The standard treatment for cardiac tamponade in leukemia is pericardiocentesis for decompression followed by radiation and steroids. We performed pericardiotomy in our patient because her cardiac function deteriorated rapidly as manifested by left heart failure. The desirability of obtaining tissue diagnosis was a secondary but important consideration in performing an open thoracotomy.

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