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Cardiac tamponade complicating leukaemia: immediate chemotherapy or pericardiocentesis?

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Summary: Although leukaemic infiltration of the pericardium is frequently observed at post-mortem, clinically evident cardiac tamponade is rare. Two cases of cardiac tamponade complicating leukaemia are presented. One patient had cardiac tamponade as the initial presentation of acute lymphoblastic leukaemia and experienced complete resolution of the pericardial effusion within 6 days after chemotherapy without therapeutic pericardiocentesis. The other patient with chronic myeloid leukaemia developed cardiac tamponade requiring pericardiocentesis as the first sign of acute blastic transformation. The roles of early chemotherapy and pericardiocentesis in managing this complication are discussed.

Introduction

Leukaemia involves all organs and tissues of the body. Leukaemic infiltration of the pericardium has been documented frequently at post-mortem examinations. Clinically, however, pericardial effusion with cardiac tamponade is rare and only isolated case reports have been described. In all the reported cases, therapeutic pericardiocentesis was required for relief of cardiac tamponade with the risk of bleeding since these patients often had deranged haemostasis. This report details two patients with leukaemia presenting with cardiac tamponade. To our knowledge, the rapid resolution of cardiac tamponade induced by chemotherapy alone in our first patient and the development of cardiac tamponade as the first sign leukaemia (CML) in the second patient have not been previously described. The role of early chemotherapy and pericardiocentesis will be discussed.

Case reports

Case 1

A 45 year old previously healthy Chinese female was admitted for fever and cough for 2 weeks as well as retrosternal chest pain which worsened on deep inspiration for 3 days. She also noticed breathlessness on exertion on the day of admission. On examination, the patient was pale, not in respiratory distress and running a fever of 37.8°C. The heart rate was 120 beats/min with evidence of pulsus paradoxus and the blood pressure was 100/50 mmHg. The jugular venous pressure was elevated up to the angle of the jaw. The cardiac apex was ill-defined and heart sounds were faint. There was no murmur or pericardial friction rub. Abdominal examination revealed 4 cm hepatomegaly and a spleen tip. Chest radiograph revealed globular cardiac silhouette and a right pleural effusion. Electrocardiogram (ECG) showed generalized diminished voltage and sinus tachycardia. An echocardiogram revealed a large pericardial effusion with gross cardiac oscillation and diastolic collapse of right ventricle and atrium. Laboratory investigations showed haemoglobin 7.3 g/dl, white blood cell (WBC) count 7.3 × 10^9/l with 79% blasts, platelet count 45 × 10^9/l. The prothrombin time and activated partial thromboplastin time were normal. Bone marrow aspirate and cytochemical and immuno-cytochemical studies confirmed the diagnosis of T-cell acute lymphocytic leukaemia (ALL). Smears on the pleural fluid aspirated with platelet infusion cover using the Gram and Ziehl-Neelsen stains, and cultures for aerobic, anaerobic, and tubercle bacilli were negative. It showed a WBC count of 12.8 × 10^9/l with 90% lymphoblasts.

She was transfused and was promptly started on systemic chemotherapy comprising vincristine, cyclophosphamide, prednisone and daunorubicin. Therapeutic pericardiocentesis was withheld owing to the relative lack of symptoms, while the patient was closely monitored with serial chest radiographs and echocardiography. There was symptomatic improvement within 48 hours with disappearance of pulsus paradoxus and normalization of jugular venous pressure. The blood pressure went up to 120/70 mmHg.
the pulse rate decreased to 90/min. Echocardiogram
2 days after chemotherapy revealed reduction in size
of pericardial effusion and absence of diastolic com-
pression of right ventricle. The temperature returned
to normal on day 4 and another echocardiogram on
day 6 revealed complete resolution of the pericardial
effusion. Chest radiograph revealed decreased cardio-
thoracic ratio and resolution of the pleural effusion.
The patient remained in haematological remission and
there was no evidence of recurrence of pericardial
effusion 6 weeks afterwards.

Case 2

A 19 year old female with Philadelphia chromosome
positive CML was maintained in the chronic phase for
the past 5 years with busulphan. She was admitted into
hospital because of rapid onset of sharp retrosternal
chest pain, epigastric pain and shortness of breath over
24 hours. Physical examination showed she had
central cyanosis and cold peripheries. The blood
pressure was 80/50 mmHg and pulse rate was 140/
min, with marked pulsus paradoxus. The jugular
venous pressure was raised and Kussmaul’s sign was
elicited. The cardiac impulse was ill-defined and the
heart sounds were distant. A typical pericardial fric-
tion rub was heard. The liver was palpable 4 cm below
the right costal margin and was tender. The spleen was
palpable 10 cm below the left costal margin. Chest
radiograph revealed cardiomegaly. Echocardiogram
revealed sinus tachycardia and diffuse non-spe-
cific T wave inversion. Echocardiogram confirmed
the presence of pericardial effusion with diastolic
collapse of right ventricle and right atrium. Haemo-
globin was 10.8 g/dl, platelet count 126 × 10^9/L and
WBC count 39.4 × 10^9/L with blasts 15%. Cyto-
chemical and immunophenotyping studies failed to
define the lineage of the blast cells.

In view of the severity of her symptoms, pericar-
diocentesis was performed via the subxiphoid approach.
Two hundred millilitres of straw-coloured fluid was
obtained and the patient’s clinical status improved
dramatically. Analysis of the pericardial fluid showed
protein 55 g/L, glucose 7.8 mmol/L, WBC count
5.6 × 10^9/L with 40% blast cells. Cultures for bacteria,
fungus and acid-fast bacilli were repeatedly negative.
Bone marrow trephine biopsy confirmed CML in
blastic transformation. She was given hydroxyurea
with initial response. The WBC count decreased to
13.7 × 10^9/L. Echocardiogram showed a residual rim
of pericardial effusion.

Four weeks later, however, she was readmitted
again because of epigastric pain, shortness of breath as
well as generalized erythematous plaque-like lesions
over the face and trunk. Another chest radiograph and
echocardiogram showed recurrence of massive
pericardial effusion. The WBC count rose to
41.4 × 10^9/L with 39% blasts. In view of the recurrent
nature of her pericardial effusion, pericardotomy with
drainage was performed; 375 ml of blood-stained fluid
was released. Pericardial biopsy revealed a thickened
fibrotic pericardium with perivascular infiltration of
inflammatory cells and fibroblasts. There was no
definite evidence of leukemic infiltration nor infec-
tion. Biopsy of the cutaneous erythematous plaque
showed marked leukemic infiltration of the dermis.
She was given chemotherapy comprising vincristine,
cytosine arabinoside and thioguanine. There was no
reaccumulation of pericardial effusion and the pericar-
dial drain was removed after 4 days. However, she died
4 weeks later because of fulminant pneumonia. Post-
mortem examination was refused.

Discussion

In a large autopsy study of 420 patients with acute
leukaemia, Roberts et al. observed leukemic
infiltration in the hearts of 156 patients (37%), 99 of
whom showed pericardial infiltrates. However, it is
unusual for the cardiac involvement to be manifested
clinically and even rarer to be the initial presenta-
tion of leukaemia.

Chu et al. in 1983 reviewed the literature and found
17 reported patients with leukaemia who had manifesta-
tions of pericarditis with cardiac tamponade. They
included 9 patients with ALL, 5 with acute myeloblas-
tic leukaemia, 2 with CML and one with chronic
lymphocytic leukaemia. Cassis et al. described a
45 year old man with massive haemopericardium as
the initial manifestation of CML. One similar patient
with ALL was described by Mancuso et al. Review of
all these cases showed that all patients required
pericardiocentesis in addition to chemotherapy and/or
radiotherapy for control of the pericardial effusion.

The first patient in this report showed several
interesting features. First, the cardiac tamponade was
the initial presentation of her ALL. Second, the blast
cells in this patient were of T-cell origin. T-cell ALL
with cardiac tamponade has also been described by
Mancuso et al. The cell types in other reported cases
of ALL with cardiac tamponade were not specified.
T-cell ALL is well known to have mediastinal involve-
ment, but whether T-cell ALL has a predilection for
pericardial infiltration remains to be seen. Third, the
pericardial effusion responded dramatically to
chemotherapy, with complete resolution within
6 days. Such rapid resolution of cardiac tamponade in
leukaemia induced by chemotherapy alone has not
been previously reported.

Cardiac tamponade complicating CML is espe-
cially rare, and occurred in the chronic phase of the disease
in all the reported cases. Its occurrence as the first
clinical manifestation of acute blastic transformation,

as in our seco previously.

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previously.
All the evidence points towards malignant
infiltration as the cause of the pericardial effusion in
our patients. In the first patient, the pericardial
effusion resolved rapidly with chemotherapy, and the
coexistent pleural effusion was proven to be leukemic
in origin. In the second patient, the pericardial fluid
showed blast cells, and was repeatedly culture
negative. The accompanying extensive leukemic
infiltration of the skin further supported this.
Moreover, the recurrence of the effusion correlated
with the activity of the blastic stage of the disease.
A negative pericardial biopsy does not disprove the
diagnosis, as leukemic pericardial infiltration may be
scattered.1

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