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AN UNUSUAL PRESENTATION OF ACUTE LYMPHOBLASTIC LEUKAEMIA WITH PERI-CARDIAL EFFUSION CAUSING CARDIAC TAMPONADE
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SUMMARY

Peri-cardial effusion is most commonly associated with tuberculous infection in the developing world. Peri-cardial effusion causes symptoms when it is large or when it has accumulated rapidly. Non-tuberculous causes of peri-cardial effusion include bacterial infections, uraemia, viral infections, rheumatic fever, connective tissue disorders, post – peri-cardiotomy syndromes and malignancy. We present a case of acute lymphoblastic leukaemia first presenting as a large peri-cardial effusion causing tamponade.

INTRODUCTION

Leukaemia is a systemic disorder involving all organs and tissues of the body. Cardiac involvement is seen at necropsy in approximately 20% of of patients with malignant lymphomas or leukaemias (1). However, clinically, peri-cardial effusion with cardiac tamponade is rare and only isolated case reports have been described (2-4). lt is very unusual for acute leukaemia to present as cardiac tamponade without any clinical or haematological evidence of the disease. We describe a patient with acute lymphoblastic leukaemia showing this most unusual mode of clinical presentation.

CASE REPORT

Seven year female patient presented with a one week history of cough, easy fatiguability and progressive dyspnoea. Three months prior to this the patient had been treated for recurrent upper respiratory infections. There was no positive history of contact with an adult with a chronic cough. There was no history of evening fevers, drenching night sweats or marked weight loss. Childhood Immunisations were up to date according to national programme schedule. Her Milestones had been achieved normally.

Clinical examination revealed a sick-looking girl who was anxious and markedly dyspnoic. She had warm peripheries, regular peripheral pulses, with a rate of 120 beats/minutes. Her jugular venous pressure was elevated to the level of the jaw and her blood pressure was 90/50mmHg. The cardiac apex was displaced, heart sounds were faint with no added sounds. The chest was clear with normal quality breath sounds. The abdomen was mildly distended, with a tender haepatomegaly of four centimetres below the costal margins. There was no splenomegally, nor any other masses. An impression of severe bronchopneumonia with congestive cardiac failure with a differential diagnosis of a large peri-cardial effusion was made.

Laboratory investigations revealed a white blood cell count (WBC) of 4.12×109/l, haemoglobin 9.1g/dl, a platelet 268×109/L and an erythrocyte sedimentation rate of 10 mm/hour. A peripheral blood film showed no abnormal cells with a differential count of 78% lymphocytes, 18% neutrophils, 3% monocytes and 1% eosinophils. An HIV test was negative, mantoux test was non reactive, Ziehl-Neelson (ZN) stain on sputum revealed no acid fast bacilli (AAFB). Urea and electrolytes and liver function tests were normal. Blood cultures were negative for bacteria.

The chest X-ray showed massive cardiomegaly. An echocardiogram revealed a large peri-cardial effusion. Emergency peri-cardiocentesis was done and 400 ml straw coloured fluid was tapped. The fluid showed glucose levels of 2.0mmol/L, and a protein concentration of 48g/L. Gram and ZN stains were negative for bacteria and AAFBs respectively. Microscopic examination of the peri-cardial fluid showed a reactive picture as it had scattered clumps of polymorphs including eosinophils, macrophages and lymphoid cells Mantoux test was non – reactive.

She was treated with broad spectrum antibiotics, prednisone, Lasix and digoxin. The patient improved markedly while in the ward and was discharged subsequently, to be followed up in the clinic. She was seen again a month later with cough associated with difficulty in breathing and easy fatiguability with no fever. On examination she was found to have cervical and submandibular lymphadenopathy. She had a pulse rate of 116 beats/mins which was of normal volume and the

heart sounds were normal. A chest X-ray revealed a normal sized heart with hilar nodes. An echo-cardiogram showed no peri-cardial effusion. The haemoglobin was 14.8g/dl, WBC of 86.1 x 10^9/L and a platelet count of 146 x 109/l. A peripheral blood film revealed neutrophils 4%, stabs 5%, lympocytes 14% , eosinophils 1% and *blasts/ atypicals 76%*, described as atypical mononuclear cells many having morphologic characteristics of blasts. The ESR was 37mm/hour. The child was re – admitted with a diagnosis of acute leukaemia. A bone marrow aspirate showed features of acute lymphoblastic leukaemia (ALL-L2). Cytology of the cerespinal fluid revealed abundant small abnormal lymphoid cells. She was promptly started on systemic induction chemotherapy comprising of vincristine (2mg/m^2), adriamycin (60mg/m^2). In addition she received intrathecal methotrexate alternating with Cytarabine (10 doses in total) and oral Prednisolone15 mg TDS. Unfortunately she did not go into remission after the fourth cycle of induction and was started on re-induction on second line chemotherapy. She has been in and out of hospital for over one year and is in poor state. She did poorly and got systemic and CNS relapses after a year of treatment. She succumbed while undergoing third re-induction.

DISCUSSION

Peri-cardial effusion with cardiac tamponade, in acute leukaemia, is rare and only isolated case reports have been described (2-4). It was found in a large autopsy study of 420 patients with acute leukaemia, that 41 of them had leukaemic infiltrates in the peri-cardium. Roberts *et al*, observed that of 156 patients with peri-cardial effusions, 99 (63%) showed peri-cardial infiltrates (5). However, widespread literature search revealed that most papers report that it is unusual for the cardiac involvement to be manifested clinically and even rarer for it to be the initial presentation. A review of 78 cases by Muir *et al* of patients with cardiac tamponade as the initial presentation of their malignancy, only 9% originated from leukaemia or lymphoma, while lmbach found it to be 7% (6, 7). In an article published by WebMED Professional on Maligant Peri-cardial Effusion, amongst the list of causes, primary tumours come first, followed by metastatic or infiltrative diseases, under which leukaemias do not even feature (8). Baleydier *et al* found that in 41 cases of T cell ALL, the presence of pleural/peri-cardial effusion in these children was four out of eight (20%) (9). In a study of 59 patients with pleural effusion half (54%) of them had a concomitant peri-cardial effusion and in only one of them there was cardiac tamponade. In this latter case peri-cardial fluid and blood works were unremarkable. It took two more months for the leukaemia to reveal itself (11). An identical history and progression was later reported by Chia *et al* in 1973, (12). In this case we have presented, peri-cardial effusion did not reoccur most likely because of prednisone which was incorporated in the initial treatment after she presented with tamponade.

In conclusion, we report a very unusual case of acute lymphoblastic leukaemia first presenting with cardiac tamponade without any clinical or haematological evidence of the disease.

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