Case report

Juvenile rheumatoid arthritis with cardiac tamponade

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SUMMARY A 4-year-old girl with seronegative systemic juvenile rheumatoid arthritis developed acute cardiac tamponade. Pericardiocentesis and systemic corticosteroids resulted in complete recovery of the pericardial involvement. This was followed by complete remission of rheumatoid disease.

Pericarditis with effusion has been shown to be common in juvenile rheumatoid arthritis (Bernstein *et al.*, 1974). It is usually mild and may be asymptomatic (Bernstein *et al.*, 1974; Lietman and Bywaters, 1963). However, it very rarely leads to cardiac tamponade. We believe that this is the second reported case of juvenile rheumatoid arthritis with cardiac tamponade successfully treated by pericardiocentesis (Scharf *et al.*, 1976).

Case report

A Kuwaiti Arab child, born November 1972, developed juvenile rheumatoid arthritis at the age of 2½ years that required admission to Sabah Hospital, Kuwait (May 1975). Features of her disease included intermittent fever, symmetrical arthritis of both wrists and knee joints, and severe morning stiffness. Aspirin relieved her symptoms, but the disease remained mildly active with persistence of joint swellings till her second admission in February 1976. She then had to be readmitted to the Chest Hospital with a one-week history of mild cough and dyspnoea, following 2 months of pyrexia, morning stiffness, and flare-up of arthritis.

Examination showed an ill-looking child with pyrexia and painful swelling of both wrists and knee joints. She was tachypnoeic and mildly dyspnoeic. Heart rate 120/min, regular, no murmurs or pericardial rubs heard. Chest was clinically normal. The liver was palpable 1 cm below the right costal margin with no splenomegaly. Investigations ruled out infective and neoplastic causes of her fever and showed leucocytosis and ESR 90 mm/1st h. Tests for rheumatoid factor and LE cells were negative. Chest

x-ray showed normal cardiac shadow with no evidence of pneumonia. Electrocardiogram (ECG) was normal. Aspirin for one month with serum salicylic acid levels between 18 and 43 mg/100 ml (1.3-3.1 mmol/l) was ineffective. The characteristic evanescent rash was noted on a few occasions. There was progressive hepatomegaly and dyspnoea; meanwhile the spleen became palpable and generalised lymphadenopathy was noted.

A month after readmission the child looked very ill, dyspnoeic and tachypnoeic, with intermittent grunting and mild intercostal recession. Neck veins were distended. The liver was palpable 7 cm and spleen 2 cm below the costal margin. There was no ascites, ankle or sacral oedema. Heart rate was 150/min, respiratory rate 44/min, blood pressure 80/50 mmHg. Chest x-ray showed marked cardiac enlargement without pulmonary congestion (Fig. 1). ECG showed generalised nonspecific ST changes and diminished QRS amplitude in most leads. Echocardiography confirmed pericardial effusion.

Emergency cardiac catheterisation and cardioangiography proved the presence of tamponade. Haemodynamic studies proved systemic congestion (right atrial mean pressure 12 torrs) with 'M' shaped pattern of the right atrial pressure curve. Cardioangiography after the injection of the contrast dye into the right atrium (15 ml 76% Urografin, 10 ml/s) showed a wide distance between the right atrial cavity and the right heart border (Fig. 2).

Pericardiocentesis was performed. The RPX polyethylene catheter 045 (Beckton-Dickinson) was introduced into the pericardial cavity by the Seldinger technique (Erben *et al.*, 1970) and 140 ml of sero-sanguinous fluid was removed. Total proteins of the fluid were 59 g/l, sugar 10 mg/100 ml (0.555 mmol/l) (blood sugar not checked). Direct smear and culture

Fig. 1 Chest x-ray showing marked enlargement of cardiac shadow without pulmonary congestion.

Fig. 2 Angiogram showing the wide distance between the right atrial cavity and right heart border.

showed no organisms. No acid-fast bacilli were detected by Antiformin and culture was sterile weeks later. Tests for rheumatoid factor and LE cells were negative.

Systemic corticosteroids, digoxin, and frusemide were started on the same day and aspirin stopped. The following day the patient looked better and much more comfortable, but still with mild dyspnoea. Heart rate 120/min, respiratory rate 28/min, BP 100/60 mmHg. Liver was 3 cm below the costal margin and the spleen was just felt. Clinical improvement was steady and progressive; 2 weeks later the child looked happy, afebrile with no dyspnoea. Neck veins were not distended, liver was 1 cm below the costal margin with no splenomegaly; the heart on x-ray reverted to normal size (Fig. 3).

Treatment of cardiac failure was stopped one month and prednisone 2 months after pericardial centesis and the child was maintained on aspirin for 3 months. Both wrists and knee joints were normal with full range of movement. When last seen in June 1977 (off aspirin for 10 months) the child looked healthy with normal weight, and height. ESR was normal. There were no signs of constrictive pericarditis.

Discussion

Rheumatoid arthritis (RA) associated pericarditis is usually mild and may be asymptomatic. This was

Fig. 3 Chest x-ray taken 2 weeks after the first showing normal cardiac size.

shown by the low incidence described in several clinical studies (Lietman and Bywaters, 1963; Gordon *et al.*, 1973), in spite of the high incidence reported at necropsy (Young and Schwedel, 1944). Recent echocardiographic studies (Bacon and Gibson, 1972; Bernstein *et al.*, 1974) have confirmed the common association and mild nature of pericardial involvement in RA and its juvenile variant. Nevertheless, it can lead to cardiac compression though this seems to be rare. In 1975 Thadani *et al.* reviewed the literature and found only 18 well documented cases of cardiac tamponade complicating RA; to this they added 2 cases of their own. Of these 20 cases, only one was a child (originally reported by Nadas and Levy in 1961), showing that tamponade is even rarer in JRA than in RA.

Because of the lack of a specific diagnostic test for rheumatoid pericarditis, the possibility of viral pericarditis occurring in a child with JRA can present a difficult diagnostic problem. Viral pericarditis frequently follows an upper respiratory infection with a short latent period, onset is acute, and recurrences are common (Cayler *et al.*, 1963). The study and characterisation of rheumatoid pericardial fluid have provided valuable diagnostic help. Diminished concentrations of glucose in rheumatoid pleural fluid (Carr and Mayne, 1962) and rheumatoid pericardial fluid (Latham, 1966) in the absence of acid-fast bacilli and bacteria have been considered very suggestive of rheumatoid aetiology. The pericardial

fluid was studied in 8 of the 17 patients reviewed by Franco *et al.* (1972), 'decreased levels of sugar and increased levels of gammaglobulins and lactic dehydrogenase were characteristic'. Our patient, in addition to one week of mild cough, had fever and flare-up of arthritis for 2 months before admission. The sugar content of her pericardial fluid was very low.

The role of systemic corticosteroids in the treatment of rheumatoid pericarditis has not been firmly established. Nadas and Levy (1961) were reluctant to use them routinely, while Franco *et al.* (1972) believed that steroids would predictably reverse signs and symptoms of rheumatoid pericarditis. We believe that a controlled study is needed; the recent impressive echocardiographic findings should make this possible.

Mainly because of its rarity, the best method of managing pericardial tamponade is difficult to decide. Thadani *et al.* (1975) recommended pericardial resection because of the risk of fluid being thick and loculated; making it difficult to remove. We believe that this obstacle could, at least in most patients, be overcome by insertion of a catheter into the pericardial cavity.

This method adds only a little inconvenience to the routine technique of pericardial tapping and the wide diameter of the catheter should make the removal of even thick fluid much easier. In our experience this technique is safe in adults (Erben

et al., 1970), and in children the procedure is better done under general anaesthesia, preferably ketamine. We believe that pericardiocentesis should be tried first in the management of rheumatoid cardiac tamponade, while pericardectomy is being considered.

Ball *et al.* (1975) described the presence of gamma-globulin complexes in rheumatoid pericardial fluid and the absence of these complexes in the patient's serum, suggesting the local production of these complexes. Scharf *et al.* (1976) injected corticosteroids locally into the pericardium in addition to performing pericardiocentesis, and the improvement in their patient was dramatic. Our patient showed the same dramatic improvement without local corticosteroid injections, thus making it difficult to judge their value.

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