Rheumatic Valvulitis and Constrictive Pericarditis

Report of Case

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SUMMARY

A 13-year-old girl was admitted with congestive heart failure, edema, ascites, and jaundice. There was an apical pansystolic murmur of mitral insufficiency and marked cardiomegaly. Her venous pressure was elevated. Despite medical treatment her condition deteriorated, hepatic and renal failure as well as disseminated intravascular coagulation ensued, leading to her death.

At post mortem she was found to have rheumatic mitral valvulitis and constrictive pericarditis. The pathologic picture of pericarditis was nonspecific, but in presence of a positive skin test for tuberculosis the latter is considered to be the most likely cause of the pericarditis, nevertheless, rheumatic etiology of pericarditis in this case cannot be excluded. The presence of rheumatic heart disease and cardiomegaly may have led to the exacerbation of symptoms and signs of constrictive pericarditis and severe right heart failure.

Additional Indexing Words:
Disseminated intravascular coagulation Pericarditis Pulsus paradoxus Rheumatic fever Tuberculous pericarditis

Although rheumatic fever is considered to be the most common cause of pericarditis in childhood, it is generally stated that it seldom if ever leads to constrictive pericarditis.

Tuberculosis has been considered as the most common cause of constrictive or adhesive pericarditis, nevertheless, the number of cases labeled as idiopathic form exceeds those with a known etiology. To our knowledge the combination of rheumatic valvulitis and adhesive pericarditis has not been reported. The purpose of this communication is to report a patient with rheumatic mitral insufficiency and constrictive pericarditis presenting a virulent course of myocardial, hepatic and renal failure. The possibility of rheumatic etiology of the constrictive pericarditis is entertained.

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CASE REPORT

A 13-year-old girl was admitted to the Ahari Children's Hospital Medical Center, because of generalized edema, dyspnea ascites, and jaundice. Dyspnea and edema had appeared 6 months prior to admission. During the 3-month period preceding the admission the abdomen had gradually enlarged and jaundice appeared.

The child had been in good health up to 11 years of age, when she had been at another hospital with a diagnosis of acute rheumatic carditis. She was found to have a pansystolic murmur at the apex, and the liver edge had been 4 cm below the right costal margin. Her chest roentgenogram had shown marked cardiomegaly.

Her antistreptolysin O titer (ASOT) had been 833 Todd units and the erythrocyte sedimentation rate (ESR) 34 mm. The Mantoux test had been positive. She had been discharged in fair condition after 1 month. She had not received regular prophylaxis and had progressively become dyspneic. Physical examination at the time of admission showed a 13-year-old girl in acute distress. She had obvious jaundice, her pulse rate was 140/min, and respiration was 48/min. She weighed 32 Kg and her blood pressure was 135/80 mmHg. The peripheral pulses were of fair quality and pulsus paradoxus was evident. There was 2+ pitting edema of lower extremities. The neck veins were distended and inspiratory engorgement was evident (Kussumal sign). A left precordial bulge was present. The apical impulse was rather heaving and diffuse, felt at the 6-7th left intercostal spaces outside the mid-clavicular line. The first heart sound was muffled, the second heart sound had physiologic splitting and a protodiastolic gallop rhythm was audible. A grade III/VI high pitched apical pansystolic murmur with radiation to left axilla as well as a low pitched middiastolic rumble were heard. The liver was hard and 3–6 cm below the right costal margin, there was ascites, and 2+ generalized edema.

Fig. 1. Chest roentgenogram AP view and barium swallow, lateral view.
Note the marked cardiomegaly, and left atrial enlargement.
Chest roentgenogram showed marked cardiac enlargement (Fig. 1) and pulmonary congestion. A barium swallow showed left atrial enlargement.

An electrocardiogram showed an axis of +80°, the PR interval was 0.24, and there was evidence of biatrial enlargement and flattening of the T waves over most of leads. The tracing was also suggestive of left ventricular hypertrophy, evidenced by a ventricular activation time of 0.06 in V₆, but the R voltage in V₆ was 8 mV. The hemoglobin was 16.3 Gm/100 ml and hematocrit 49%. The white blood cell count was 7,500/cumm with 82% neutrophils, and toxic granulations were noted. The ESR was 5 mm, ASOT 1,250 Todd units and C-reactive protein 1+ positive. The urinalysis was negative except for a trace of albumin. The serum bilirubin was 11 mg/100 ml with 6.9 mg/100 ml direct fraction. Serum transaminases (SGOT and SGPT) were 105 and 43 Karmen units respectively. Prothrombin time was 52% of normal. Total protein was 6.6 Gm/100 ml with an albumin of 3.5 Gm/100 ml. The A/G ratio was 1:1, the protein electrophoretic pattern was normal, and thymol turbidity was 1.6 units. Blood glucose was 143 mg/100 ml. Alkaline phosphatase was 12 King-Armstrong units. Serum electrolytes were normal. The central venous pressure was found to be 28 cm of water. The patient was placed on digitalis, penicillin, and diuretic (Furosemide). Despite the therapy, the patient's condition was deteriorating. During the second week of admission the blood urea nitrogen was 32 mg/100 ml and the total serum bilirubin had gone up to 38 mg/100 ml. She was oliguric; at this time the child developed generalized purpura, the signs of heart failure were persistent and the degree of edema and ascites had increased. The platelet count was 20,000/cumm, the bleeding time 13 min, clotting time more than 1 hour, and prothrombin time 90 sec with a control of 13 sec. With a diagnosis of disseminated intravascular coagulation the patient was started on in-

Fig. 2. A close up photograph of the heart. Note the thickening and adhesion of pericardial layers.
travenous heparin therapy. The general condition continued to deteriorate and she developed hematemesis. The hemoglobin dropped to 11 mg/100 ml and SGOT and SGPT 180 and 66 units, respectively. A repeat and third series of coagulation tests showed persistent and increasing abnormalities, she received blood transfusion as well as anticongestive therapy, heparin, and massive antibiotic therapy. But succumbed 14 days after admission.

At autopsy the heart weighed 500 Gm, there was extensive fibrosis of the pericardium with shaggy fibrinous and thickened fibrous bands covering both ventricular chambers encompassing the vascular roots and atrio-ventricular junctions, and there was adhesion of pericardial layers as well as severe adhesions to the entire heart (Fig. 2). The mitral leaflets showed moderate thickening of the free border with focal nodosities. No calcifications were present. The chordae tendineae were slightly shortened, the papillary muscles were moderately hypertrophied, and the main pulmonary vessels were free of emboli. On microscopic examination there was moderate hypertrophy of the myocardial fibers with perivascular hyalinization and infiltration of mononuclear cells some of which were larger myocytic cells. Anitschkow myocytes and early Aschoff body formations were seen in many parts of myocardium (Fig. 3). The pericardium showed large amounts of fibrinous material admixed with fibrous tissue and large numbers of mononuclear cells. No stigma of tuberculous lesions were found, and acid fast statining and culture for tuberculous bacilli were negative (Fig. 4).
DISCUSSION

The presence of rheumatic mitral insufficiency and congestive heart failure at the time of admission was beyond any doubt, and there was no evidence in favour of any congenital cardiac anomalies.

Certain features, however, could not be well explained on the basis of mitral insufficiency, namely the presence of ascites, increased venous pressure, and jaundice. The liver tests, although somewhat abnormal, were not in favour of any primary hepatic disease causing the jaundice and ascites. It was thus felt that the jaundice could perhaps result from some extrahepatic biliary obstruction, which in turn has also caused venous obstruction and ascites. This however was refuted by an essentially normal level of alkaline phosphatase. The possibility of constrictive pericarditis was entertained, and subsequently supported by increased venous pressure, an enlarged liver and ascites. The heart size in constrictive pericarditis is generally mildly to moderately enlarged and marked cardiomegaly is somewhat unusual. In the presence of severe mitral insufficiency that was present in our case we would expect severe left ventricular hypertrophy as well. The positive evidence in favour of left ventricular hypertrophy in our case was a VAT of 0.06 sec, and although the voltages were normal in view of the expected left ventricular hypertrophy the electrocardiogram can be said to have low voltage. The presence of flat T waves is also in favour of chronic pericarditis. Despite the prevalence and severity of rheumatic heart disease in Iran, we considered it extremely unusual for a child with rheumatic mitral insufficiency to develop right heart failure and liver function derangement at such an early age. This feeling was supported by the absence of clinical findings of pulmonary hypertension or right ventricular hypertrophy. Thus although not quite certain we felt that the most probable explanation for the patient's clinical picture would be a combination of rheumatic mitral insufficiency and constrictive pericarditis as well as cardiomyopathy. Certain clinical findings such as pulsus paradoxus, a positive Kussmaul sign, elevated venous pressure, ascites and hepatomegaly support this view. We feel that the association of pericarditis and severe cardiomegaly secondary to mitral valve disease and myocarditis may have created a clinical and hemodynamic state indistinguishable from constrictive pericarditis. The etiology of pericarditis in our case is not certain.

Aside from tuberculosis, viral infections have also been felt to lead to this condition. The fact that a tuberculous pericarditis can be negative for tuberculosis on histological and microbiological grounds is well documented. The association of rheumatic carditis and tuberculous pericarditis
has not been reported. The anamnestic background in this case with its evolution and the valvular lesions makes the diagnosis of rheumatic carditis a certainty. The pericardial lesion, however, remains an enigma. The preponderance of tuberculosis in this part of the world makes it most probable etiological agent. According to certain statistics only 30% of cases of constrictive pericarditis are proven to be tuberculous by culture and animal inoculation. Others, however, believe that many cases labelled as idiopathic constrictive pericarditis are in fact of tuberculous etiology.

Although to our knowledge the association of tuberculous adhesive pericarditis and the rheumatic endocarditis has not been reported, we believe this to be largely due to decreasing incidence of both of these maladies in western hemisphere.

In this part of the world, however, both systemic tuberculosis and rheumatic heart disease are far from rarity.

The possibility of rheumatic pericarditis leading to chronic adhesion in this case can be entertained particularly in view of the absence of evidence in favour of any other pathologic process.

The myocardial failure in the present case can be related to rheumatic lesions, since the subpericardial damage in myocardium in itself was not enough to explain the extent of heart failure manifested clinically.

It is doubtful if surgical intervention and pericardiectomy would have been fruitful in this case since the myocardial and valvular involvements were extremely severe.

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