The Turkish Journal of Pediatrics
Volume 27, No. 4, October - December 1985

A CASE OF CONSTRICTIVE PERICARDITIS IN AN INFANT TREATED BY RADICAL PERICARDIECTOMY*

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Key words : constrictive pericarditis, infant, pericardiectomy, tubular pericarditis

Constrictive pericarditis complicating purulent pericarditis is unusual in infants, but the possibility of it should be born in mind as if it goes unrecognized the prognosis is very poor[1-4]. The disease usually follows a chronic course characterized by ascites, shortness of breath and failure to thrive. The symptoms may be confused with those of primary liver or kidney disease. In survey reports of pericarditis, the incidence of chronic constrictive pericarditis in children ranges from 1% to 13% of all cases[5]. Thus, although it is rare in the childhood age group, it yet occurs frequently enough for a knowledge of its clinical presentation, cause and hemodynamic abnormalities to be important. The following case report documents the rapid development of constrictive pericarditis in a four-month-old infant and emphasizes both the importance of early diagnosis by noninvasive tests and cardiac catheterization, as well as treatment by pericardiectomy.

Case Report

A four-month-old boy had been treated with streptomycin, penicillin G and trimethoprim-sulfamethoxazole for five days for a painless swelling seated at the posterior cervical triangle. The swelling decreased in size, but in the meanwhile the child developed shortness of breath and fatique, so his family took him to the Erciyes University Hospital. There physical examination revealed tachypnea,

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tachycardia, faint heart sounds, hepatomegaly, and edema of the hands and feet. The chest X-ray demonstrated cardiomegaly and paracardiac infiltration (Figure 1). An ECG demonstrated elevation of the ST segment. Needle aspiration of the

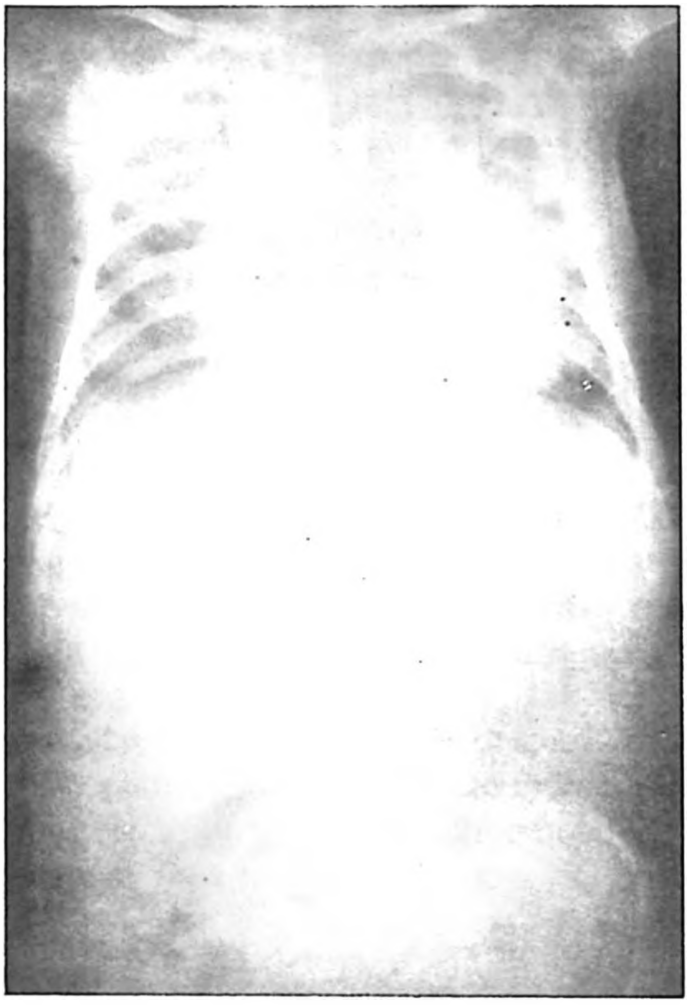
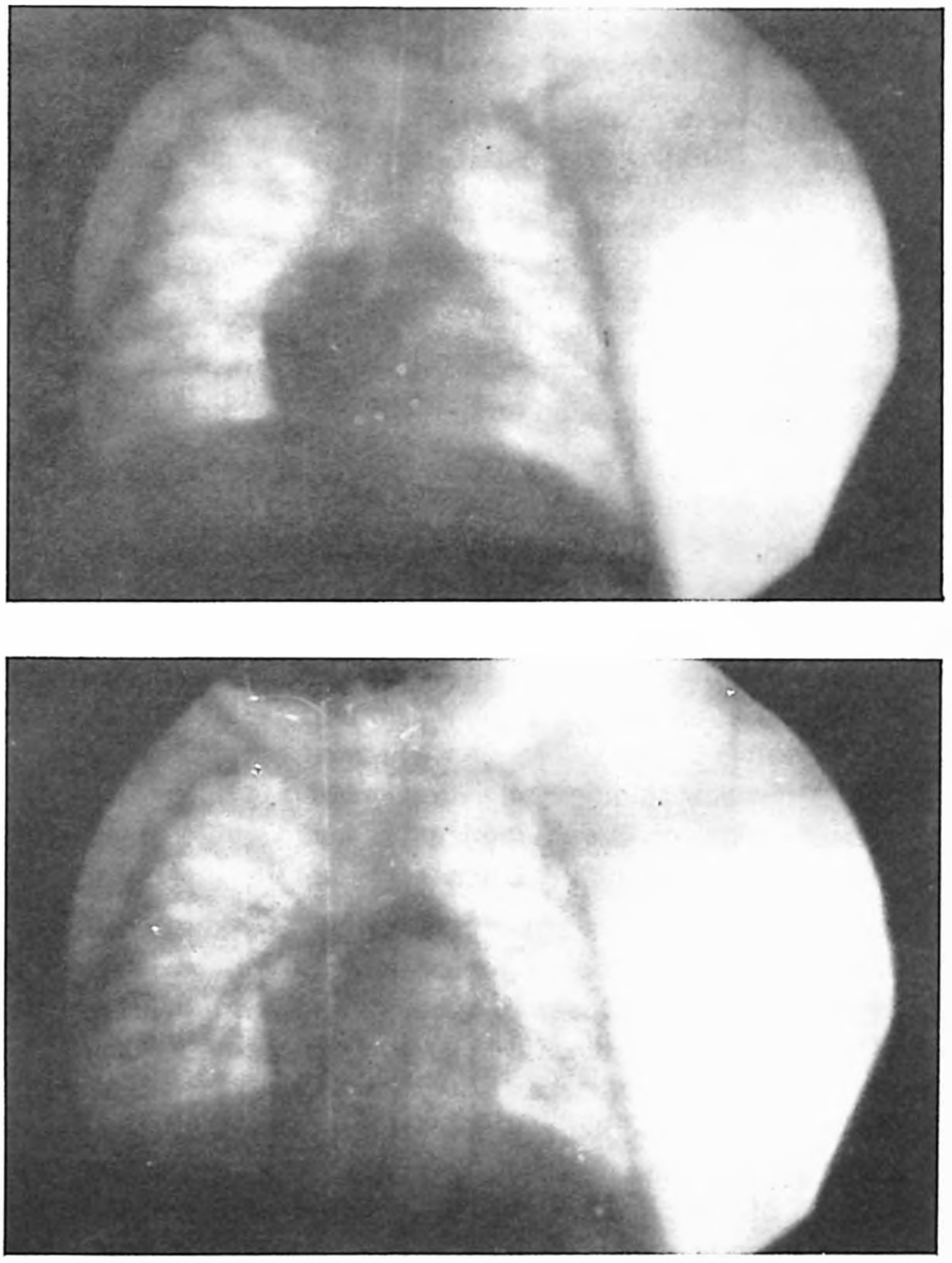


Figure 1
A telecardiogram showing preoperative global cardiomegaly.

pericardial cavity revealed Staphylococcus aureus and therapy with prednisone, gentamycin, and methicillin was begun. Since, however, there was no satisfactory clinical improvement the patient was transferred to our hospital where he was digitalized, and in addition to the antibiotics initially given, furosemide was also begun. The echocardiogram demonstrated a thickened pericardial wall without effusion, and since there was a clinical diagnosis of pericardial constriction, cardiac catheterization was performed. The data from cardiac catheterization revealed a right atrium mean pressure of 13 mmHg, a right ventricle pressure of 29/16 mmHg, and a pulmonary artery pressure of 30/10 mmHg. The right atrial angiograms demonstrated straightening and thickening of the right ventricular border and medial displacement of the left ventricle by a thick peel around it (Figures 2,3).



Figures 2, 3

Right atrial angiograms showing straightening and thickening of the right cardiac border and superior vena caval dilatation.

A median sternotomy was performed and the anterior mediastenum was entered. There were white necrotic plaques on the thymus. The heart was found to be contricted by a thick peel within the pericardial sac, and this severely restricted its motion. The pericardium and peel were dissected off the anterior and diaphragmatic surface of the heart. The peel was approximately five mm thick and was severely adherent to the epicardium. There was no evidence of acute infection at the time of surgery. The pathological examination revealed fibrinous pericarditis

and granulomatous inflammation with Langhans type giant cells in the thymus and surrounding tissues.

Though the skin tests were negative and pericardiocentesis did not yield acid-fast bacilli pathological examination suggested the presence of tuberculous pericarditis with fibrinous pericarditis which was probably secondary to acute purulent staphylococcal pericarditis.

After the operation the patient did well for four days, and the cardiac symptoms improved dramatically; but on the fifth day he developed fatique, diarrhea, and tachycardia, and on the following he died of sepsis. The last lomber puncture culture revealed Staphylococcus aureus.

Discussion

Constrictive pericarditis is an uncommon problem, especially in infancy. Since mortality continues to be high in this age group, it is imperative to recognize the entity and institute therapy as soon as possible. The clinical picture is characterized by the triad of a small heart, ascites and elevated venous pressure[1-6]. However, as in our patient, constrictive pericarditis frequently presents as congestive heart failure unresponsive to medical therapy. In the infant, the subtle physical findings of a jugular venous pulse showing no giant waves, Kussmaul's sign, or even a pericardial knock are especially difficult to evaluate. Other tests such as the ECG, echocardiography, and chest X-ray may aid diagnosis.

The ECG can demonstrate biatrial enlargement, right axis deviation, right ventricular hypertrophy, and diffuse nonspecific changes in the ST segments and T waves[1-7]. This pattern of right axis deviation and right ventricular hypertrophy is believed to be secondary to heavy basilar fibrosis leading to cardiac distortion and rotation; and altering the direction of electrical forces to simulate right ventricular hypertrophy. On the X-ray, the cardiac silhouette may be globular in configuration, with a suggestion of pericardial thickening or even calcification. Further, there may also be an enlarged superior vena caval shadow, as well as pleural effusion[6].

In constrictive pericarditis several echocardiographic findings have frequently been observed, these include pericardial thickening, early pulmonary valvular opening, early interventricular septal diastolic motion, rapid E-F slope, and the flattening of the left ventricular endocardium[7]. There are, however, no specific patterns pathognomonic of constrictive disease. In our case, pericardial thickening, rapid E-F slope and early interventricular septal motion were evaluated by echocardiography.

Although these noninvasive tests provide additional information to support the diagnosis of constriction, the most definitive diagnostic procedure is cardiac catheterization[6,8]. Hemodynamic findings are the results of impaired ventricular

filling and stroke volume. The right atrial pressure tracings show no respiratory variation with the typical "M" or "W" wave form, steep X and Y descents are prominent, and late diastolic pressures in the great veins and in the atria are increased. Ventricular pressure tracings demonstrate an early diastolic dip and a late diastolic plateau, the so-called square root sign. In our patient, a right atrial angiogram demonstrated straightening and thickness of the right cardiac border and superior vena caval and also left atrial dilatation; however, with restrictive cardiomypathy there may be findings similar to those just described for constrictive pericarditis. The rapid volume expansion during cardiac catheterization results in equilibration of all diastolic pressures and might be helpful in diagnosis[8,9]. Further in distinguishing between these two diseases the presence of concentric pericardial calcium on fluoroscopy of chest X-ray is a helpful sign, though it may not be present in all cases of constrictive pericarditis, especially in childhood.

After the diagnosis of constrictive pericarditis has been made, treatment, consisting of fluid restriction, digitalis and vigorous diuresis is instituted. The poor prognosis of patients who are not operated on, underlines the necessity for urgent radical pericardiectomy[10]. The operation can be performed through a bilateral anterior thoracotomy incision with division of the sternum. However, we prefer a median sternotomy incision. This affords excellent exposure to the pericardium and allows a wide resection of the pericardium, including removal of the pericardium from both ventricular surfaces, atria, and vena cavae. Frequently, as in our case, it becomes necessary also to excise a thin layer of the epicardium which is often involved in the constrictive process. Radical pericardiectomy has resulted in markedly improved survival rates in children with constrictive pericarditis. The determinant factors in survival and improvement appear to be the chronicity of the disease and the degree of underlying myocardial fibrosis.

In the department of thoracic and cardiovascular surgery of our hospital, pericardiectomy has been applied in 67 cases of pericarditis between 1968 and 1983. Of these surgical interventions 30 were carried out after a short-term medical follow-up and the other 37 of them after a long-term medical follow up. The medium age was 18.9. Only in three cases (approximately 4.5%) was the patient under one year of age. The reason for pericardiectomy in this group was purulent pericarditis due to staphylococci and viruses. In our case, the pathological diagnosis was compatible with tuberculosis and in this respect it would seem to be the only case of its kind for this age group[5,8]. The morbidity and mortality rates for pericardiectomy which has been carried out after the appearance of pericardial constriction, or in the late phases of long-term medical follow-up, have usually been found to be higher than if the operation has been carried out at an earlier phase[11-13]. To prevent irreversible myocardial and hepatic changes due to long-term constriction, it is often recommended that the pericardiectomy be

carried out during the short-term medical follow-up, that is three to six weeks from the beginning of pericarditis, and before there are extensive adhesions, fibrosis, and calcification[10,12,14]. We are fully in accord with this view.

Summary

A four-month-old boy with constrictive pericarditis, which is unusual in infancy, is presented. A radical pericardiectomy was performed on the patient. It is of interest that pathological examination of the resected pericardium revealed the presence of tuberculous pericarditis with fibrinous pericarditis, this was probably secondary to acute purulent staphylococcal pericarditis.

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