Recurrent Idiopathic Pericarditis: Failure of Corticosteroid Therapy*

Report of a Case

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IDIOPATHIC PERICARDITIS IS BEING RECOGNIZED WITH INCREASING FREQUENCY. Most cases recovered uneventfully with rest and symptomatic treatment. A few patients require corticosteroid therapy for relief of symptoms. In rare instances even the most intensive medical management fails, and the patient develops chronic relapsing illness characterized by recurring chest pain, fever, weakness and dyspnea. Zinsser and co-workers were the first to draw attention to this syndrome, which results in prolonged disability, at times amounting to virtual invalidism.

CASE REPORT

A 23-year-old white housewife was first examined in February, 1958 because of anterior chest pain of two days' duration. There had been four previous attacks of pain during the preceding 14 months, the first occurring when she was three months pregnant. These episodes were mild and persisted for only a few days. She had received no treatment.

The present attack came on suddenly without preceding respiratory infection or other evidence of illness. She denied fever, cough, malaise or dysphagia. The pain was well localized to the upper anterior chest; it was aggravated by deep inspiration, twisting of the trunk, or lying supine. She was most comfortable in a semirecumbent position. She denied having dyspnea. There had been no swelling of the neck, face or feet.

On physical examination, she was afebrile, pulse 66, regular; blood pressure 122/68; weight 144 pounds, stripped. There was no distention of the neck veins when she lay supine. Findings on examination of the heart and lungs were unremarkable. On fluoroscopy of the chest, no abnormality was noted except questionable dilatation of the superior vena cava.

Laboratory studies showed a hemoglobin of 11.9 grams, PCV 37 per cent, sedimentation rate 35 mm./hr., and WBC 12,200 (N 72, L 28). Intradermal testing with intermediate strength PPD, coccidioidin 1:100 and histoplasm 1:100 was uniformly negative. Posteroanterior and lateral x-ray films of the chest were within normal

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Figure 1: P.A. and lateral chest films at the height of the patient's illness showing pericardial effusion and bilateral pleural effusion.

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limits and a barium swallow was negative.

Early on the morning of the fourth day of illness, the pain suddenly became much more intense and she was immediately admitted to the hospital in acute distress. On examination at this time, she appeared pale and in severe pain. Oral temperature 99.0° F., pulse 84, regular; blood pressure 114/87. The neck veins were distended in the upright position. She was unable to lie down even at an angle of 45°. A chest x-ray film showed definite enlargement of the cardiac shadow, both to the right and to the left (Fig. 1).

Pericardial aspiration was performed through the fourth left anterior interspace just adjacent to the sternal margin. Two hundred fifty ml. of greenish turbid fluid was aspirated with immediate relief of pain, and 200 ml. of air was instilled into the pericardial sac, demonstrating thickening of the pericardium (Fig. 2). The pericardial fluid was negative on culture for routine organisms, tubercle bacilli, and fungi. No abnormal cell was seen in the spun sediment.

Following pericardial aspiration, she was begun on penicillin-streptomycin injections with some improvement in fever and leukocytosis for the next four days. Electrocardiogram was normal at this time. Second strength PPD was negative on two different occasions. Two L.E. preparations were negative.

On the eighth day of illness, there was a rise in temperature and an increase in chest pain, orthopnea, and malaise. The liver, for the first time, became palpable 4 cm. below the right costal margin, and moderate pitting edema of the feet and sacrum was noted. A triphasic friction rub was also heard for the first time. Pericardiocentesis was repeated and only 60 ml. of bloody fluid was obtained. There was little relief of symptoms following this aspiration. Venous pressure measured 14 cm. of water. The following day her fever was 102° F., pulse 130, and the friction rub had disappeared. Prednisolone, 40 mg. daily, was started with immediate fall in temperature and improvement of all symptoms.

Because of the dramatic response to corticosteroid therapy, consideration of surgery was deferred and she was discharged from the hospital. Prednisolone was continued at home, along with oral penicillin, and later chloramphenicol. Electrocardiogram at this time showed inverted T waves in leads II, III, AVF, and V1 through V6 compatible with the diagnosis of pericarditis.
For six weeks she continued to do well, but then chest pain, fever, dyspnea and weakness returned. She gained weight rapidly and developed acne, hypertension, moonface, and peripheral edema as concomitants of the hyperadrenocortical state.

She was readmitted to the hospital and prepared for surgery by careful regulation of her electrolyte balance and corticosteroid dosage. In addition, she was placed on prophylactic streptomycin-isoniazid. On May 25, 1958, three and one-half months after the onset of the fourth recurrence of her pericarditis, a bilateral anterior thoracotomy was carried out. At surgery, the parietal pericardium was found to be thickened, varying from 3 mm. to 10 mm. and could be removed without excessive difficulty. The heart appeared grossly normal and there were only 20 to 30 ml. of clear fluid within the pericardial sac. No tumor or granuloma was found. She had a smooth postoperative course and corticosteroid therapy was tapered off over a period of two months, when she resumed normal activity.

Since operation four years ago, this woman has had another baby after normal pregnancy and delivery. There has been no evidence at any time of recurrent or persistent pericarditis. Serial chest films and ECG's have remained normal (Fig. 3).

**DISCUSSION**

This case presents several noteworthy features. The patient was a young woman, whereas most reported cases have been in men, in a ratio exceeding 3 to 1. The etiology was never established even after careful study of the excised pericardial tissue. Tuberculosis, fungus diseases, and collagen diseases were ruled out. Four minor episodes occurred during a 14-month period, culminating in a severe, unremitting illness. The relapsing course is similar to that described by Zinsser in a small group of patients.

We wish to emphasize the failure of corticosteroid therapy to control this patient's symptoms even when given in large amount over a prolonged period. Despite vigorous medical treatment, she became disabled to the point of invalidism. Her disease was controlled and good health restored only after pericardiectomy was carried out. At the time of this report, four years after pericardiectomy, she remains in good health and free of any evidence of persisting or recurrent disease.

**REFERENCES**