SELECTED REPORTS

Carcinoid Tumor of the Thymus Associated With Recurrent Pericarditis*

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A 49-year-old man with carcinoid tumor of the thymus presented with the clinical picture of acute pericarditis. Recurrent bouts persisted after removal of the tumor without other evidence to indicate tumor recurrence. There were no associated manifestations of an endocrine disorder.

Thymic tumors are a common cause of anterior mediastinal masses. A most uncommon thymic tumor, a carcinoid, is described here associated with recurrent pericarditis.

CASE REPORT

A 49-year-old man was found to have a round, anterior mediastinal mass on a chest x-ray film taken on Feb 23, 1977. A month earlier, he had experienced an episode of right shoulder pain radiating to the upper sternal region, which was made worse by deep breathing and which subsided spontaneously.

Roentgenographic examination in March 1977 showed no change. Arch aortography and tomography added no new information. Tomograms failed to show calcium within the tumor, and the visualized surrounding structures appeared normal.

The patient was not seen again until late August 1977, when he was admitted to the University of Alberta Hospital with symptoms of acute pericarditis. He had severe substernal pleuritic chest pain and fever. On examination his temperature was 38.2°C. Blood pressure was 120/80 mm Hg. No pericardial rub was heard, and physical examination was otherwise normal. Treatment with indomethacin resulted in complete remission of signs and symptoms.

Laboratory investigation disclosed a white blood cell count of 10,900/cu mm on admission, increasing to 17,600/cu mm on the third hospital day. Erythrocyte sedimentation rate (ESR) reached a maximum of 29 mm/hr. SGOT was increased to 128 (normal, 40 to 50), and lactic dehydrogenase (LDH) to 383 (normal, <200). LDH fractionation showed an increase in liver or skeletal muscle fraction. Both enzymes returned to normal with remission of symptoms. An ECG on admission was normal, but the following day there were typical changes of acute pericarditis. The tracings returned to normal six days after admission. Echocardiogram failed to show evidence of pericardial effusion. Chest x-ray film again demonstrated the mediastinal mass (Fig 1).

An extensive investigation undertaken to determine the etiology of the pericarditis was unrewarding. This included viral studies, Mantoux test, and studies for histoplasmosis and toxoplasmosis.

On Sept 8, 1977, the patient underwent median sternotomy. A tumor measuring approximately 7.0 × 7.0 × 7.5 cm was apparent in the right anterior mediastinum. The apparently well-encapsulated tumor was free of the lung, the tumor was adherent to the anterior pericardium adjacent to the great vessels. Minimal serous pericardial fluid was present, which was later found to be sterile and contained no malignant cells. A complete excision of the mass was possible along with wide margins of adjacent pericardium. His early postoperative course was uneventful.

Pathologic examination revealed a well-encapsulated tu-

Figure 1. PA chest x-ray film on admission. Right superior hilar mass.

Figure 2. Photomicrograph showing carcinoid tumor composed of relatively uniform polygonal cells arranged in ribbons and rosettes. No mitotic activity. Supporting stroma is delicate fibrous trabeculae (× 250).

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tumor with focal areas of hemorrhage. Under light microscopy the histologic features of a carcinoid were noted, with uniform tumor cells arranged in ribbons and rosettes (Fig 2). Mitotic activity was minimal. Extensive necrosis and hemorrhage were apparent throughout the specimen. The attached pericardium did not show evidence of invasion.

Electron microscopy confirmed the presence of neuroendocrine cells of the Kulchitsky type (Fig 3). Cytoplasmic dense core granules were noted within most of the tumor cells. This, then, was believed to be a well-differentiated thymic carcinoid.

Following his discharge from the hospital, the patient did well until early November 1977, when he was readmitted with recurrent pericarditis. He was given indomethacin, and the symptoms subsided within three days. Since then, the patient has had at least three other bouts of pericarditis in February, April, and May 1978. During each of these attacks there was general malaise, fever, leukocytosis, and elevated ESR. During the most severe of these attacks, in May 1978, there was radiologic evidence of pericardial as well as a small pleural effusion. There was also elevation of alkaline phosphatase and SGOT. Therapy was begun with a maintenance dose of indomethacin for six months, and he has remained asymptomatic and free of tumor since that time.

**DISCUSSION**

Carcinoid tumors occur uncommonly in the thymus. They may or may not be associated with Cushing’s syndrome, with the syndrome of multiple endocrine adenomatosis, and with other systemic manifestations. As with other thymic tumors, they may be clinically silent and found by a routine chest x-ray film, or they may present with symptoms of local invasion.

These tumors tend to be more aggressive than thymomas, but prolonged survival after local invasion and distal spread is not uncommon. Treatment has usually consisted of excision of the tumor, sometimes followed by local radiation. Following surgical excision more than 50 percent of the patients without endocrine abnormalities may be expected to survive without evidence of tumor. Recurrences and lymph node metastases appear to be most common in those patients who had grossly invasive tumor at the time of the operation. Generalized metastases are rare.

The patient described here did not show clinical evidence of an associated endocrine disorder or systemic manifestations, except for those of recurrent acute pericarditis. To our knowledge, this has not been previously reported, and the nature of this association in our patient remains obscure.

Initially, it was thought that the most likely explanation was pericardial invasion by the tumor, although certain atypical features were recognized, such as the prominent systemic manifestations, the elevation of serum enzymes of possible liver origin and the prompt clinical response to indomethacin. At the time of surgery, however, the tumor was found to be well-encapsulated, with only very local pericardial invasion.

Subsequent bouts of pericarditis have also been self-limited, between periods of complete good health and without evidence of tumor recurrences. The clinical features of each attack have been those of acute nonspecific pericarditis and the possibility of a coincidental association cannot be excluded here.

The etiology of this condition has often been thought to be viral but, in one study, supportive evidence could only be found in five of 34 patients. It may be recurrent in about 20 percent of the cases. It is not clear whether these recurrences are due to persistence of the causative agent or to the setting up of an "autoimmune" mechanism, such as has been postulated for the postpericardiotomy syndrome. Any of these factors may have been operative in our patient; however, the most likely explanation is coincidental viral pericarditis.

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**REFERENCES**