Adrenocortical Carcinoma with a Giant Pericardial Mass

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A patient with a history of adrenocortical carcinoma presented with massive pericardial effusion and a giant pericardial mass. Death resulted from cardiac tamponade. At autopsy a large necrotic mass was histologically similar to the original adrenocortical carcinoma. We present this case and review the literature on metastatic pericardial disease.
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Key words: cardiac tamponade, pericardial metastasis, CT scan, Mitotane

Introduction

Although pericardial metastases are not unusual, pericardial masses are rarely encountered(1–3). We present a rare case of adrenocortical carcinoma in which cardiac tamponade and a giant mass in the pericardial cavity developed, and review the literature on metastatic epicardial masses.

Case Report

A 53-year-old woman was admitted to the hospital on May 30, 1991, with increasing dyspnea and progressive cardiomegaly. In 1988, she had undergone left adrenalectomy and nephrectomy for a non-functioning adrenocortical carcinoma with metastasis to the left lung. Postoperatively she received chemotherapy (Mitotane 1,500 mg/day) with no change in size of the lung mass.

On admission, the blood pressure was 122/62 mmHg and the heart rate was 108/min, with a paradoxical pulse. There were no rales in the lung fields. Heart sounds were normal without murmurs or a pericardial friction rub. Facial edema was evident but there was no leg edema.

Chest radiographs revealed marked cardiac enlargement and a mass in the left middle lung field. A 12-lead electrocardiogram showed sinus tachycardia, depressed ST segments and inverted T waves in leads I, II, III, aVF and V4–6. Echocardiography disclosed a solid mass and a large effusion in the pericardial cavity. Computed tomography (CT scan) of the chest disclosed an enhancing mass in the pericardial cavity (Fig. 1A). Metastasis of adrenocortical carcinoma to the epicardium was suspected. The right atrial mean pressure was 18 mmHg, and the mean pulmonary capillary wedge pressure was 21 mmHg. The cardiac index was 1.64 L/min/m².

Laboratory data showed a leukocyte count of 10,400, C-

reactive protein of 9.1 mg/dl, and an erythrocyte sedimentation rate of 60 mm/hr. In addition, a slight elevation in transaminase probably due to passive liver congestion was found. Neuron-specific enolase was as high as 66.2 ng/ml. The urinary 17-hydroxycorticosteroid and 17-ketosteroid were normal. The pericardial fluid was bloody with a protein concentration of 5g/dl and was characterized as an exudate.

Surgical drainage of the pericardial cavity was performed on June 2 and a total of 2,000 cc was evacuated. Doxycycline and minocycline were then infused into the pericardial cavity. When drainage stopped, the pericardial tube was removed. No increase in pericardial effusion was subsequently observed.

The pericardial mass increased its size within 3 months, with the CT scan demonstrating an area of low density in the mass, which was considered to reflect necrosis (Fig. 1B). The paradoxical pulse recurred on July 29 and the low cardiac output decreased further. The patient died on August 25.

Autopsy revealed no local recurrence of adrenocortical carcinoma, but multiple metastatic foci were found in the lungs. The heart weighed 1,300 gm. The space between the epicardium and the myocardium was infiltrated with necrotic tumor (Fig. 2A). The histologic picture of the mass was consistent with the previous adrenocortical carcinoma, with typical large clear cells and multiple mitotic figures (Fig. 2B). The pericardial mass was, therefore, diagnosed as a metastatic adrenal cortical carcinoma.

Discussion

Cardiac metastasis of a malignant tumor is not rare, with a reported incidence of 9.7–12.2% (4). Malignant tumors which commonly metastasize to the heart include lung cancer, breast cancer, malignant lymphoma, leukemia and hypernephroma. The pericardium is most common site of cardiac metastasis

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Fig. 1. Computerized tomographic scan with contrast medium on admission demonstrating massive pericardial effusion and a solid mass (arrow) in the pericardium. A nodule (arrowhead) is also noted in the left lung (A). Three months later, the pericardial mass had become enlarged, resulting in an area of low density in the pericardium (B).

followed in frequency by the myocardium. Pericardial effusion is frequently an accompanying occurrence of pericardial metastasis (5). If it is advanced, cardiac tamponade ensues, requiring drainage or pericardiocentesis. In rare cases, epicardial metastases produce a pathologic state similar to constrictive pericarditis (4). However there have been no prior reports of such a giant pericardial mass demonstrated radiologically (1–3, 6, 7).

Adrenocortical carcinoma is a relatively rare malignant tumor, with a frequency of 0.0002% (1, 8). Furthermore, non-functioning adrenocortical carcinoma is said to be less frequent than the secretory type. This may account for the paucity of

Fig. 2. Section of the heart demonstrating a giant mass (arrow) in the pericardium with a necrotic component (A). Histologic picture reveals clear cells with multiple mitotic figures resembling the adrenocortical carcinoma removed previously (B) (x400).

reports of cardiac metastasis of adrenocortical carcinoma. Adrenocortical carcinoma is a highly malignant tumor (2). From 30% to 50% of patients present with metastases at diagnosis, and in only 4–7% is the tumor confined to the adrenal gland (3, 9).

In the present case, based on the existence of mediastinal lymph node metastasis, a lymphatic route was suspected. Although the mechanism of formation of a giant tumor mass in the pericardial cavity is unclear, some relationship to such high malignancy is suspected.

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