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CARDIOLOGY
Case Report

Malignant Solitary Fibrous Sarcoma Presenting as Primary Heart Tumor

Fu-Chung Chen*

Division of Cardiovascular Center, Department of Medicine, Lin Shin Hospital, Taichung, Taiwan

***Corresponding Author:** Fu-Chung Chen, Division of Cardiovascular Center, Department of Medicine, Lin Shin Hospital, Taichung, Taiwan.

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Abstract

This paper describes the case of a 42-year-old male patient with primary heart malignant solitary fibrous sarcoma presenting with cardiac tamponade. The patient received urgent surgery and adjunctive chemotherapy nevertheless he died 14 months later. To our knowledge, this is the first described case of a primary cardiac tumor causing cardiac tamponade to the point of needing emergency decompression surgery.

Keywords: *Heart tumor; Cardiac tamponade; Surgery*

Case Report

A 42-year-old male patient presented at our emergency department with progressive exertional dyspnea, chest tightness and chest pain and near syncope. There was a rapid irregular pulse, blood pressure 94/62 mm Hg, bilateral low lung moist rales and muffled heart sounds and pulsus paradoxus. A chest X-ray showed an increase in the size of the heart and redistribution of lung markings consistent with interstitial pulmonary edema. The initial diagnosis was of congestive heart failure, and an inotropic agent and a low-dose diuretic were prescribed. Progressively-worsening shock and heart collapse followed, subsequent to which echocardiography (Figure 1) showed a large mass surrounding the myocardium, causing tamponade of the left ventricle. A multiple-slice computed tomographic (MSCT) scan (Figure 2) showed a huge heterogeneous mass that was causing compression of the left ventricle, but the coronary arteries were observed to be normal. Emergency surgery was performed to relieve the left ventricle compression. The entire pericardium cavity was found to be filled with a soft mass, which was diffusely infiltrating the left ventricle wall. A curative resection was deemed impossible and debulking was performed. The tissue pathology confirmed the presence of a malignant solitary fibrous sarcoma (Figure 3).

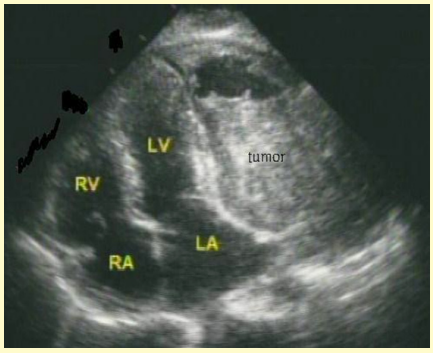

Figure 1: *An apical two-chamber echocardiograph showed a huge mass with central necrosis inside the pericardium, causing tamponade of the left ventricle (LV: left ventricle; LA: left atrium; AO: aortic outlet).*

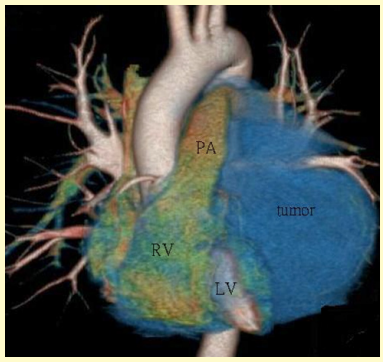

Figure 2: *64-slice CT showed a huge heterogeneous mass of about 15 cm in length with hematoma formation and contrast enhancement in the posterior heart portion inside the pericardium, causing left-ventricle compression (LV: left ventricle).*

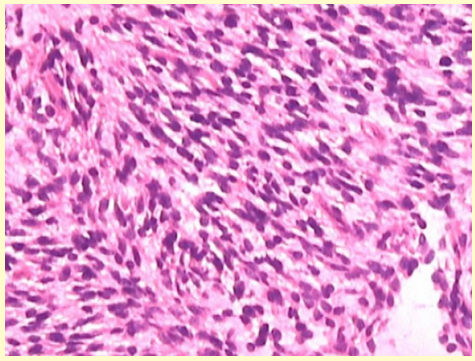

Figure 3: *The microscopic histological picture indicates hypercellular lesions demonstrating cytologically atypical medium-sized round to spindle tumor cells. Immunohistochemical staining revealing the tumor cells to be positive for Bcl2 and negative for CD117, CD 34, desmin, actin, EMA, keratin and S100.*

The patient received adjunctive chemotherapy and radiotherapy but still died 14 months later owing to multiple lung metastases.

Discussion

We report here an unusual case of a large malignant solitary fibrous tumor in the pericardium causing acute cardiac tamponade. In threatened cardiac tamponade, management should be directed towards urgent pericardiocentesis because of right ventricle compression caused by fluid accumulation, but this method was unsuitable in our case of soft tissue compression. When urgent tamponade is diagnosed, intravenous hydration should be instituted, especially in patients who mistakenly received a diuretic owing to an incorrect diagnosis of congestive heart failure, as in this case.

Solitary fibrous tumor of the pleura was first described by Klemperer and Rabin in 1931. It is a rare neoplasm derived from mesenchymal tissue. Since then, there have been a series of reviews of solitary fibrous tumor of the pleura (SFTP) describing the clinicopathologic features of the neoplasm [1]. More than half of all patients with such tumors are asymptomatic, and 80% of such masses are benign [2-4]. Also, cardiac sarcomas are uncommon tumors [4]. Approximately 25% of primary cardiac tumors are malignant, and of these about 75% are sarcomas. In a large reported series, angiosarcoma was the most common (37%), followed by malignant fibrous histiocytoma (MFH, 24%), leiomyosarcoma (9%), rhabdomyosarcoma (7%), unclassified (7%), and others (16%). The prognosis for malignant

solitary fibrous tumor is much more guarded [5]. Approximately 63% of patients will have a recurrence of their tumor, of which more than half will succumb to disease progression within 2 years. Adjuvant chemotherapy and radiotherapy in malignant solitary fibrous tumor remains controversial. Pathological examination demonstrated the tumor to be composed of increased mitotic activity and cellularity of spindle cells with a collagenous matrix [4]. Immunohistochemical staining was positive for CD34 and Bcl-2 and negative for CD31. Our patient showed positive for Bcl-2 and negative for CD34 [6]. Extrinsic compression of this type of solitary fibrous sarcoma may cause patients to seek medical care, with the common symptoms of cough, dyspnea, chest pain and even syncope, as in our patient. The size of the tumor can vary greatly—between 1 cm and 36 cm (mean, 6 cm) in diameter [7,8]. Owing to its large size, limitation inside pericardium and direct compression of the left ventricle causing cardiac tamponade, complete surgical resection of the tumor can present many challenges, particularly given its proximity to vital neighboring structures [7]. Such tumors have never been reported as being of cardiac origin. In the case of our patient, when the pericardium was surgically opened, the tumor was protruding out, and we are not very clear about the origin of the tumor. There is a possibility that it might have originated inside the pericardium. A primary tumor is defined by the original site (organ or tissue) at which the tumor started. A primary heart tumor is a better result than a secondary one. We considered the tumor as having a primary origin of the heart for two reasons: the first is imaging modalities showing the tumor only inside the pericardium and free of the peri-aorta lymph node or other organs, and the second is that the primary tumor site is usually larger than the secondary site. To our knowledge, this is the first described case of a malignant solitary fibrous sarcoma causing cardiac tamponade to the point of needing emergency decompression surgery.

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