Clinical-pathologic conference in cardiac surgery: Malignant schwannoma of the heart

Tomaso Bottio, MD, PhD
Gino Gerosa, MD
From the University of Padua,
Padua, Italy

Participants

From the University of Padua
Padua, Italy

Cardiovascular Surgery
Dr Tomaso Bottio
Dr Gino Gerosa

Cardiology
Dr Roberta Montisci

Radiology
Dr Francesco Corbetti

Pathology
Dr Vito Ninfo

Case Presentation

Dr Bottio: The patient was a 51-year-old man presenting to a local emergency department in January 2003 with sinus node tachycardia, chest pain, and dyspnea. Laboratory data and arterial blood gas analyses were within normal values. The electrocardiogram demonstrated tachycardia (105 beats/min), left ventricular hypertrophy, and Q-waves in the anterior leads that were hypertrophy related. He had a chest radiograph that demonstrated a mediastinal abnormality with pleuropericardial effusion. Cardiac physical examination was unremarkable, as was his medical history. The hematologic and biochemical values were within normal limits. Transthoracic echocardiography, computed tomography (CT), and magnetic resonance imaging (MRI) were performed. Perhaps we could ask Dr Montisci to comment on transthoracic echocardiography.

Dr Montisci: Two-dimensional echocardiography showed a mildly enlarged aortic root (38 mm). Doppler scanning showed normal cardiac valve apparatuses. The left atrial chamber was compressed by an intrapericardial mass, without signs of discontinuity from the myocardial wall. The left and right ventricular chambers were normal in size and function. The right atrium was encircled and compressed by the mass, and the superior and inferior venae cavae were dilated. Only the right superior pulmonary vena cava was detectable at echocardiography.

Dr Bottio: Dr Corbetti, would you describe CT and MRI?

Dr Corbetti: The CT scan revealed an extensive mass growing into the pericardium and left thorax. The MRI is remarkable for an intrapericardial mass, probably originating from the pericardium. It is mostly homogeneous, except for the presence of some small cystic areas, and it grows deeply in the pericardial cavity at the back of the heart. The major diameter of the mass is 18 cm. The heart is dislocated up and forward (Figure 1). The heart is totally encircled by the mass; the left and right atrial chambers are compressed, and the pulmonary veins are tightened (Figure 2). Apparently the myocardium is not infiltrated by the mass. At MRI scanning, the inferior pulmonary lobe of the left lung is atelectatic, and a left pleural effusion is also present; also, the inferior vena cava is encapsulated (Figure 3). There are mildly enlarged (8- to 9-mm) lymph nodes in the peritracheal, tracheobronchial, and periaortic positions.

Dr Bottio: A percutaneous needle biopsy of the mass was performed to characterize the mass. Dr Ninfo, would you describe the histology of the lesion?

Dr Ninfo: The histology showed fusiform cells with evident signs of several mitoses distributed within areas with different grades of cellularity. The first hypothesis was for a malignant lesion from peripheral nerve sheaths.

Dr Bottio: The patient underwent an extensive workup that included a total body CT scan, the results of which were unremarkable, and a skeleton scintigraphy, which showed signs of hypercaptation within the body of the sternum. He also underwent an esophageal gastric duodenoscopy, which showed an extrinsic compression at the

From the Departments of Cardiovascular Surgery, Cardiology, Radiology, and Pathology, University of Padua, Padua, Italy.

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Address for reprints: Tomaso Bottio, MD, Institute of Cardiovascular Surgery, Via Giustiniani, 2, 35128 Padova, Italy (E-mail: tomaso.bottio@unipd.it).

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Figure 1. Sagittal MRI showing the tumor mass (§) compressing the heart. # Left ventricle; * right ventricular outflow; °aorta.

medium third of the esophagus. Bronchoscopy revealed compression and stenosis of the left main bronchus. The CT scan and MRI of the brain were unremarkable. Angiography showed increased mean right atrial pressure (16 mm Hg) and a systolic pulmonary pressure of 46 mm Hg. The right pulmonary artery was enlarged, whereas the left was stenosed. The superior left pulmonary vein was not detectable, whereas the inferior was compressed. Both the caval veins were dilated. The coronary arteries were normal, without the appearance of atherosclerotic disease.

Dr Gerosa: At the time of diagnosis, the tumor was widespread over the heart and within the pericardium, and therefore curative excision was unexpected. The major intent of our therapy was a palliation of symptoms relieving the congestive heart failure, with perhaps a prolongation of survival. Our aim was also to complete the surgical excision with radiation therapy, chemotherapy, or both. At the time of the operation, the tumor was found to be completely welded to the myocardium because the margins of excision could not be identified (Figure 4). It was extended toward the left pulmonary veins, involving extensively the anterior wall of the heart. A wide excision extending to the entry of the left pulmonary veins was accomplished, but a large part of the posterior mass was unresectable. The patient had a complicated postoperative intensive care unit stay requiring a high dose of inotropic medications and intra-aortic balloon pump support.

Figure 2. Axial CT view showing the tumor mass (§) compressing the left bronchus and pulmonary veins. * Left and right bronchus; # left pulmonary veins.

Dr Ninfo: Pathologic examination revealed a tumor mass of 2200 g in weight in which the cut surface was mostly solid with a grey-white appearance. Microscopically, the margins of resection were not tumor free, and it consisted of central fibrotic areas and peripheral high-density cellular areas in which several mitotic phenomena were present. Immunohistochemical stains were performed, and monoclonal antibodies against smooth muscle actin, CD34, CD56, desmin, vimentin, calretinin, and S-100 protein were applied. The tumor tested positive for CD56 and vimentin. The tumor was classified as a malignant peripheral nerve sheath sarcoma.

Dr Gerosa: In light of the definitive diagnosis and the extreme difficulties during the surgical resection, to obtain a complete resection of the tumor, we scheduled chemotherapeutic treatment soon after the operation. Unfortunately, the postoperative cardiac performance of the patient did not allow any adjunctive treatment because of the potential cardiotoxicity of both radiotherapy and chemotherapy. Therefore the patient was discharged to home to wait for cardiac recovery, when he suddenly died in the second postoperative month.

Dr Bottio: This case is indeed not so common and thereby offers the chance to highlight the difficulty of treatment of patients affected by such dismal diseases. Primary cardiac tumors are rare clinical entities, and fortunately, most are benign and resectable; among these, atrial myxoma is the most common.¹ On the other hand, malignant primary

Figure 3. Coronal magnetic resonance view showing the invasion of the left hemithorax by the mass (S) causing a left lung atelectasis (A). The heart chambers and the abdominal organs are severely compressed by the tumor. The ascending aorta appears normal, whereas the pulmonary artery trunk is enlarged. The inferior vena cava (O) is encircled by the mass. * Left atrium; # left ventricle; ^ ascending aorta; ^ Left and right lungs.

Figure 4. Intraoperative view of the tumor mass during thoracotomy. The tumor is widespread to the entire anterior wall of the heart.

tumors, mostly represented by soft tissue sarcomas of the heart and pericardium, are usually widespread at diagnosis, and because of invasion of the underlying myocardium, their surgical resection is seldom achieved; with curative excision being unexpected, the treatment is limited to be only palliative. Therefore the prognosis for patients with such cardiac tumors is poor and usually less than 12 months.² Nerve sheath neoplasms of the heart (soft-tissue sarcomas) are extremely rare, although they have been reported.³ These tumors more commonly involve adults than infants,⁴ and their proximity to target structures determines a subtle and nonspecific clinical presentation.⁵ Usually, presenting symptoms are dyspnea, pain, or shortness of breath, which are mostly related to pericardial effusion (often hemorrhagic), syncope, and/or sudden death when in the presence of tamponade, chest pain, hemoptysis, and

fever. When growing, these tumors can compress⁶ or occupy⁷ the cardiac chambers, and also, the cardiac function might worsen, leading to intractable cardiac failure. When the hepatic veins or the vena cava are obstructed, signs of systemic congestion appear. This is the motif that explains the variety of clinical signs and symptoms related to such tumors, which are often disguised as other more common systemic diseases. The objective of surgical therapy should be a complete resection, followed by cardiac reconstruction^{8,9}; this is usually achieved with benign schwannomas, which are encapsulated and well demarcated. On the contrary, with malignant lesions, a complete resection of the mass is seldom achieved. Hypothetically, orthotopic heart transplantation could allow complete resection of cardiac tumors. Therefore cardiac transplantation,¹⁰ autotransplantation when feasible,¹¹ or both have been proposed to allow the excision of cardiac malignancy. Obviously, a careful preoperative assessment is necessary to determine whether other cardiac and noncardiac structures are involved. Nevertheless, with experience with cardiac transplantation for primary cardiac neoplasms being limited, with it being difficult to determine the tumor operability preoperatively, and with worldwide organ availability being insufficient, cardiac transplantation leads to many unresolved questions, mostly focusing on the avoidance of wasting of a donor heart. However, for these reasons and because of the high risk of tumor recurrence or metastasis during treatment with immunosuppressive drugs,¹² the majority of patients affected do not undergo transplantation. Our patient was referred to us when he had advanced disease.¹³ Thus a partial excision of the cardiac tumor, followed by adjuvant radiation therapy, chemotherapy, or both, should have been a

palliative option. Unfortunately, survival was not prolonged because this patient suddenly died soon after discharge.

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