CLINICAL VIGNETTE

Cardiac Angiosarcoma

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Introduction

Primary care physicians frequently encounter chest pain in young people thought to be functional or due to common heart disorders, such as mitral valve prolapse (MVP). We present a twenty-four-year old woman with a very rare malignant cardiac tumor. We also review presentations of other adult cardiac tumors, both benign and malignant, primary and metastatic, that a primary care physician might encounter.

Case Report

The patient was a 24-year-old Caucasian female, accompanying her father who has cardiomyopathy to a routine office visit to her father's cardiologist. She unexpectedly had sustained palpitations, leading the cardiologist to perform an EKG and echocardiogram in his office. Her echo revealed a large right atrial mass with preserved bi-ventricular function. She had no history of weight loss, chest pain, fever, or dyspnea.

She underwent a surgical procedure to remove the mass with clear surgical margins. Pathology reported a primary cardiac angiosarcoma. Chemotherapy was offered, but the family declined since no residual tumor was identified. She received monthly echocardiograms for five months after the surgery at which time a new right atrial mass was identified She was again offered chemotherapy but declined due to concerns about side-effects. We continued to see her in hopes of finding some treatment that would provide better quality of life and to provide support to the patient and her family. She died after sudden cardiac arrest eleven months after her original diagnosis.

Cardiac Tumors in Adults

Although primary cardiac tumors are rare, metastatic involvement of the heart is much more common; up to 20-40 times more common in some autopsy series.[1] Several studies estimate that cancer metastatic to the heart may occur in one out of five patients dying of cancer.[2,3] Solid tumors commonly associated with cardiac metastases include malignant melanomas, lung cancer, breast cancer, soft tissue sarcomas, renal cell carcinomas, esophageal cancer, hepatocellular carcinoma, and thyroid cancer.[4] Cardiac or pericardial metastasis should be considered whenever a patient with known active malignancy develops cardiovascular symptoms.

General mechanisms of symptom production include the following:

- Embolization of portion(s) of the mass, mimicking a pulmonary embolism or cerebrovascular accident, depending on the location of the mass;
- Obstruction of circulation producing symptoms of heart failure; and
- Direct invasion of the myocardium resulting in impaired contractility, arrhythmias, heart block, or massive pericardial effusion, with or without tamponade.

The Carney Complex is an inherited autosomal dominant disorder, characterized by multiple tumors, including atrial myxomas and various endocrine tumors. Patients also have a variety of pigmentation abnormalities, including pigmented lentigines, and blue nevi on the face, neck, and trunk.[5]

Benign Tumors

Over 75% of primary cardiac tumors are benign.[3] In adults, the majority of benign lesions are myxomas, which occur more commonly in women.[3] About 35% of myxomas are friable or villous, and these tend to present with emboli.[3] Approximately 80% of myxomas originate in the left atrium; the second most common site is the right atrium. The second most common benign cardiac tumors are papillary fibroelastomas.[4] The most common clinical presentation is embolization leading to transient ischemic attack or stroke, and less commonly, sudden death.[4,6]

Other benign adult tumors include lipomas, which have a preponderance of fatty cells, easily differentiated on cardiac MRI. One half are found in the subendocardial region, and the remainder are found in the myocardium. They can cause arrhythmias, conduction deficits, and sudden death, so they are almost always excised.[7]

Mesotheliomas can occur within the pericardium, where they are usually malignant.[8] Resection is the treatment of choice, but it has a very poor prognosis. Cardiac mesothelioma, like its pulmonary equivalent, is also thought to be related to asbestos exposure. When a mesothelioma arises in the pericardium, tamponade may occur.[8] Rarely, mesothelioma may arise as a benign tumor of the atrioventricular node.[8]

Malignant tumors comprise 15% of primary cardiac tumors with cardiac angiosarcomas the most common primary cardiac malignancy with an overall incidence of less than 0.01%.[4] Several subtypes also exist, including rhabdomyosarcoma and malignant schwannoma. An association between cardiac angiosarcoma and P53 gene mutations have been described.[9]

Typically located in the right atrium, cardiac angiosarcomas may also invade the pericardial space, causing pericardial effusion and constriction.[10] Symptoms are non-specific and include fatigue, dyspnea, and chest pain with diagnosis usually made later in the disease. Prognosis is usually poor with a median survival of 6 months from diagnosis.[10] Embolic events may cause hemoptysis if right-sided or stroke if left-sided. Local extension beyond the atrium into the great veins may cause Superior Vena Cava syndrome.[10]

Echocardiography can help determine tumor location, pericardial involvement, and impact on valvular function. Cardiac magnetic resonance imaging (cMRI) may be able to define histology and response to chemotherapy, in addition to enhanced assessment of tumor extent and functional impact.[11] If a large pericardial effusion is present, cytology may be obtained from pericardial fluid although the diagnosis is usually made during surgical exploration. If definitive treatment cannot be provided by surgery, adjunct chemotherapy and radiation therapy may be useful. Cardiac auto-transplantation has been proposed as an alternative therapeutic option.[12]

Conclusion

This unfortunate woman was found to have a right atrial cardiac angiosarcoma, and was at constant risk for hemodynamic compromise or arrhythmia. She provides us a view of its natural course without treatment other than primary resection. What is also important is that one out five patients dying of cancer may have cardiac manifestation of disease. The presence of mass or effusion in the heart or pericardium should alert the clinician to consider either local or distant sources of metastases, which may lead to a primary tumor, hopefully with a better prognosis.

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