Papillary Fibroelastoma Involving the Left Ventricular Wall

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A 71-year-old white woman presented to her primary care physician for a routine visit and was found to have a new, previously undocumented cardiac murmur. A subsequent transthoracic echocardiogram revealed a 1 cm mobile mass arising from the lateral free wall of the left ventricle. Transesophageal echocardiography later confirmed these findings. The patient underwent a left ventriculotomy and excision of a .7 cm friable mass, which was later identified as a papillary fibroelastoma (PFE) by routine histopathologic studies. We present this unique case with a review of the literature.
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Key words: Papillary fibroelastoma • Cardiac tumor • Lambl's excrescence

A 71-year-old white woman with a history of alcoholism, tobacco abuse (30 pack-years), psychogenic polydipsia, and well-controlled hypertension presented to her primary care physician for a routine evaluation and was found to have a new cardiac murmur. The patient was asymptomatic and had no history of previous cardiac disease, chest pain, palpitations, shortness of breath, fever, neurologic deficits, or weight loss. No murmur was found during a previous examination in 1997. Family history was unremarkable for any history of cardiac tumors. The patient's only prescribed medication was benazepril (10 mg) for control of her hypertension.

Figure 1. *Two-dimensional echocardiographic image showing an intracardiac mass arising from the left ventricle.*

Physical examination was remarkable for an apical 4/6 systolic murmur which radiated to the axilla. This was accompanied by a precordial thrill in the same location. The patient was alert and oriented with no focal deficits.

A transesophageal echocardiogram demonstrated a pedunculated, freely mobile .7 cm mass arising from the lateral left ventricular free wall (Figure 1). At surgery, a .7 cm friable mass was removed from the lateral portion of her left ventricular cavity adjacent to the posterior medial papillary muscle. The patient did well post-operatively and recovered without any reported complications.

Pathologic Findings

Two portions of pale yellow to pink-tan soft tissue measuring in aggregate .7 cm × 0.6 cm × .3 cm were received from the operating room (Figure 2). Microscopically, the tumor was composed of multiple papillary fronds, having a central core of dense acellular collagen covered by a single layer of endothelial cells (Figure 3). The endothelial cells showed a variable appearance between the unusual flattened type and more polygonal cells. A myxomatous matrix with spindled cells was found between the collagen core and endothelium in some areas (Figure 3). The Van Gieson elastic stain demonstrated coarse elastic fibrils and fragmented granular deposits surrounding the central core (Figure 4).

Figure 2. *Gross specimen of a papillary fibroelastoma removed from the left ventricle.*

Discussion

Primary intracardiac masses are rare (< .3%), and are usually found when echocardiography is utilized to evaluate a patient for a source of embolus. Usually, transthoracic analysis is sufficient to visualize most intracardiac masses; however, transesophageal echocardiography is better for localizing and describing an abnormal finding. Most primary cardiac tumors are benign, and are usually found in the left atrium (75%), left ventricle (5%), or in multiple sites (5%).[1]

The most common benign primary cardiac tumor is myxoma, classically described as containing copious amounts of mucopolysaccharide mixed with polygonal mesenchymal elements. These tumors are typically pedunculated, gelatinous, and friable when handled.[2,3] They are non-malignant and are typically resected on an urgent basis to avoid embolic complications. The results following surgery are generally excellent.[3–7] A 95% survival rate has been reported at median 3-year follow-up.[3] However,

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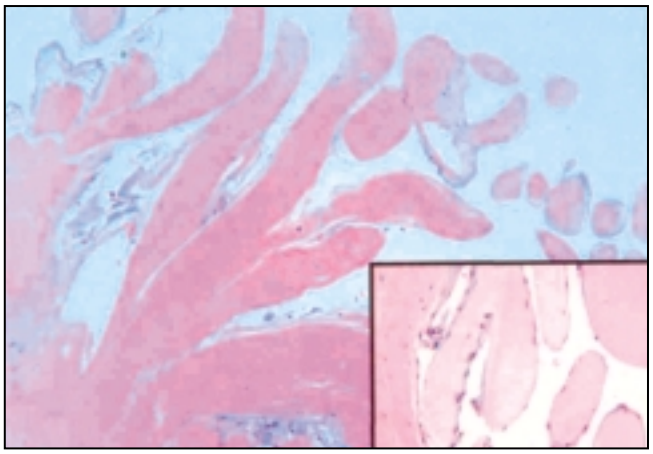

Figure 3. *(400x) Histological specimen of a myxomatous matrix between a collagen core and endothelium.*

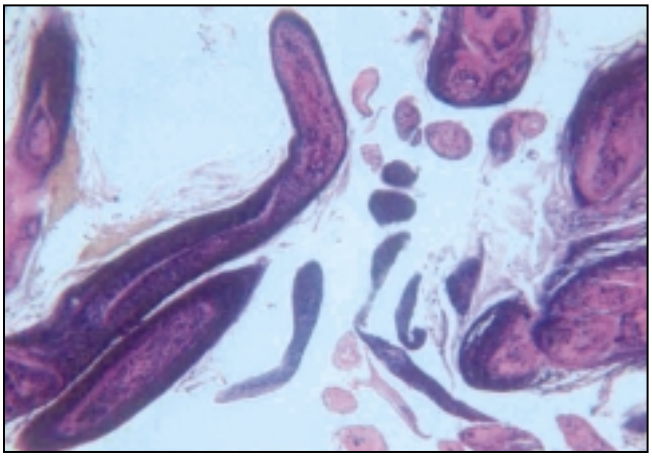

Figure 4. *(100X) Van Gieson stain demonstrating abundant concentrically arranged elastic fibers.*

approximately 5% of patients may suffer a recurrence within 5 years of treatment. Therefore, it is recommended that each patient undergo yearly echocardiography.

Papillary fibroelastoma (PFE) is a rare primary cardiac tumor that is usually found at autopsy or on echocardiography in the search for a cardioembolic source of stroke. In a study reported by the Mayo Clinic in 1992, only 7 of 110 primary heart tumors were found to be PFE.[8] Most of the 134 cases reported in the literature presented with clinical complications related to left ventricular outflow obstruction or embolic phenomenon such as acute myocardial infarction, stroke, or sudden death.[9] Numerous names have been given to this primary cardiac tumor since first being described in the 1800s. Several include papillary myxoma, fibroma, hyalofibrome, fibromyxoma, and giant Lambl's excrescence.[10]

Numerous studies have been done that describe this tumor, its rare occurrence, and the usual intracardiac locations at presentation.[8] From these data, PFE has been found to be the most common primary tumor of heart valves.[10] A study from the Armed Forces Institute of Pathology reported 39 of 45 PFEs (93%) were found on cardiac valves.[10] Similar investigations by Ryan and colleagues, and Darvishian and Farmer found 117 of 132 (89%) and 67 of 79 (84%), respectively, were primary valvular lesions.[11,12] The valves most commonly affected include the aortic (35%), mitral (29%), and tricuspid and pulmonic (10% each).[12] This tumor typically presents in adults over the age of 55, but has been described in adolescents and children as well.[10]

The underlying pathological process of the PFE is a subject currently under considerable debate. A fair number of authors believe the lesion is a reactive phenomenon resulting from persistent valvular endothelial trauma, which ultimately results in a Lambl's excrescence (LE).[13,14] Both the PFE and LE have been shown to contain fibrin and hyaluronic acid, and appear to be present along the valve edge along degeneration planes.[15]

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Unfortunately, many disagree that these two lesions are similar and point out that they differ in several ways.[16] First, LE occurs on the lines of closure, and PFE lesions are found on the smooth portions of the valve surface. LE is a relatively common

Main Points

- Papillary fibroelastoma (PFE) is a rare, benign primary cardiac tumor most commonly found in the aortic, mitral, tricuspid, and pulmonic valves.
- Patients typically present with complications related to left ventricular outflow obstruction or a thromboembolic event such as myocardial infarction or stroke.
- Primary intracardiac tumors should be resected on an urgent basis to avoid embolic complications. Survival rates following surgical intervention are excellent.
- PFE is typically found in patients over 55 years of age, but it has also been reported in adolescents and children.
- Asymptomatic patients with new cardiac murmurs should be evaluated for intracardiac tumors before significant thromboembolic events occur.

occurrence, found in 70%–80% of elderly valves, whereas the PFE lesions are extremely rare. LE are typically multiple lesions, whereas PFE are almost always single lesions. Finally, LE is reported to always contain a fibrin component, whereas PFE does not.

Primary neoplasia of the heart is very uncommon, and a high index of suspicion is needed to limit the morbidity and mortality associated with undiagnosed disease. The present case demonstrates the importance of evaluating an individual with a new cardiac murmur before significant thromboembolic or primary cardiac sequelae occur. ■

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