REVIEW

Cardiac Valvular Tumors: Cardiac Papillary Fibroelastoma

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Case histories of proposed life insurance are presented to introduce the topic of cardiac valvular tumors. Using fibroelastoma as the prototypical cardiac tumor, pathology, diagnosis, echocardiographic findings and clinical course are reviewed, based on available clinical literature. Although the natural history of benign cardiac tumors is uncertain, because of the risk of adverse outcomes, cases must be underwritten on an individual basis until long-term studies become available.

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A 47-year-old male without known risk factors for cerebrovascular disease presented with blurry vision of the left eye followed by partial visual loss. Ophthalmologic evaluation revealed the presence of ciliary retinal artery occlusion in conjunction with central retinal artery occlusion. Echocardiography revealed “small thin linear echo densities on the ventricular surface of the (aortic) valve. These densities exhibit chaotic independent motion consistent with fibrin strands.” An extensive workup revealed no other significant abnormalities, and 1 physician suggested that the ocular event “may be embolic from aortic fibrin strands.”

A 26-year-old female noted transient right-sided blindness and right tongue numbness, then a similar episode 6 months later, and a brief episode of decreased vision 7 years after. Recurrent splinter hemorrhages of the fingers have occurred. Initial workup revealed narrowing of the distal basilar artery on magnetic resonance angiography, confirmed at angiography. An echocardiogram report stated “There is an erratically moving linear echodensity (1.2 cm) attached to the right coronary cusps of the aortic valve consistent with a Lambl’s excrescence. There is probably 1 other very small (0.2 cm) Lambl’s excrescence at the coaptation of the cusps.” The report indicated that the Lambl’s excrescences were smaller compared to a study performed 3 months earlier, and previously noted mitral valve Lambl’s excrescences were no longer visible.

A 52-year-old otherwise healthy male experienced several minutes of vertigo, headache and right facial numbness. Three years previously, a brief episode of nausea and vertigo occurred while lifting weights. Magnetic resonance imaging revealed 2 abnormal signals in the posterior/inferior cerebellar hemi-
sphere felt to possibly represent subacute infarcts and a focus in the right middle cerebral peduncle, also felt to represent infarction. Workup did not reveal an underlying systemic predisposing cause, although transesophageal echocardiography reportedly revealed slight stranding of the mitral valve and slight thickening of the aortic and mitral valves.

The above case histories of proposed life insurance applicants serve as an introduction to the topic of cardiac valvular tumors, which are relatively unknown entities with unknown natural history that can cause devastating consequences.

This paper will emphasize cardiac papillary fibroelastomas (PFEs) as the prototypical valvular tumor. PFEs are histologically benign\textsuperscript{1,15,20,21,23,24} small avascular tumors, reportedly ranging in size from 1 mm to 5.7 cm\textsuperscript{29} with 99% less than 2 cm.\textsuperscript{2}

PFEs are formed from normal endocardial components, including connective tissue, smooth muscle cells, elastic fibers and fibrous tissue.\textsuperscript{1,2,11,14,17,23–25,28} Histologically PFEs are organized by a dense central core of connective tissue and covered by hyperplastic endothelial cells.\textsuperscript{4,22,23,29}

Grossly PFEs have been described as resembling a sea anemone with multiple papillary fronds extending from the central core.* The PFE is usually attached to the valvular endocardium by a short stalk,\textsuperscript{22,24,29} although they can arise from any endocardial surface.\textsuperscript{4,29}

PFEs grossly appear round, oval or irregularly shaped,\textsuperscript{2} are white to tan in color and are friable, often with adherent thrombus.\textsuperscript{28} It is likely that the friability and associated thrombi are responsible for the adverse events associated with these lesions.

The etiology of PFEs is unclear, with postulated mechanisms including neoplasm,\textsuperscript{17,26} mechanical trauma,\textsuperscript{7} inflammation,\textsuperscript{26} hamartomas,\textsuperscript{7} organized thrombus\textsuperscript{17,20} and congenital.\textsuperscript{28}

Although papillary fibroelastoma is currently the most widely accepted term, these tumors have also been referred to as cardiac papillomas, giant Lambl’s excrences, papillary endocardial tumors, papillary fibromas, cardiac papillomas, myxofibromas, fibroelastic hamartomas, papillary myxomas, fibroangiomyxomas, and others.\textsuperscript{5,7,22,28,29}

Although the great majority of PFEs reported have been attached to heart valves,\textsuperscript{2,7,14,15,21–25,28} any endocardial surface of the heart can be involved, and they have been located on all cardiac chamber walls, the left ventricular septum, papillary muscles, chordae tendinae, coronary ostia, outflow tracts and other sites.\textsuperscript{2,4,16,19,20,22,24,29}

Some authorities report the aortic valve as the most common valvular location of PFEs,\textsuperscript{2,17,20,25,28,29} others cite the mitral valve,\textsuperscript{2} and some claim that all 4 heart valves are involved with equal frequency.\textsuperscript{4,19} In children, the tricuspid valve has been reported as the most commonly involved site.\textsuperscript{25,29} Some studies report equal or nearly equal frequency of left- and right-sided PFEs;\textsuperscript{4,7} whereas, others cite greater reported frequency of left-sided lesions.\textsuperscript{22,28} The reported incidence of the right-sided tumors, suggests some investigators, may reflect the relative lack of complications and infrequency of surgical removal.\textsuperscript{2}

There is divided opinion as to which surface of semilunar valves PFEs predominantly face, with some quoting the ventricular,\textsuperscript{25,29} some citing the arterial surface,\textsuperscript{2} and others maintaining that the incidence is equal.\textsuperscript{4,23} There is more general agreement that the atrial surface predominates in atrioventricular valve PFEs,\textsuperscript{2,19,23,25,29} with only 1 source favoring the ventricular surface.\textsuperscript{24}

PFEs are rarely encountered, with various studies reporting an autopsy incidence of 0.002% to 0.33%,\textsuperscript{4,5,15–17,19,22,24,25,28,29} PFEs reportedly consist of less than 1% to 10% of all encountered primary cardiac tumors, which are themselves rare entities.\textsuperscript{1,5,7,14,15,17,19,20–25,28,29} PFEs are the most common cardiac valvular tumor\textsuperscript{1,16,22,24,25,28,29} and the third most common primary cardiac tumor after myxoma and lipoma.\textsuperscript{4,7,14}

PFEs are felt to occur with equal frequency in each sex\textsuperscript{4,17,21,29} and have a wide reported age distribution, ranging from neonate to 92
years of age, although most affected patients seem to be middle aged or older, with a mean age of about 60. They are usually solitary lesions, although multiple lesions do occur rarely, with 8 lesions reported in 1 person.

Prior to the advent of echocardiography, PFEs, which were first recognized in the 19th century, were rarely encountered either at autopsy or incidentally during cardiac surgery. The first non-autopsy description was reportedly in 1981. Recent articles indicated that only 198 cases had been reported in the literature.

Although initially felt to be asymptomatic and of no clinical significance, cardiac PFEs are now felt to be the cause of serious embolic events resulting in such clinical sequelae as cerebrovascular accidents, angina, myocardial infarction, sudden death, splenic infarctions, right ventricular outflow tract obstruction, pulmonary emboli, transient ischemic attacks, congestive heart failure, syncope, seizures and retinal artery emboli. Platelet and fibrin accumulation has been documented on PFEs, which are felt to be able to embolize. Fragments of tumor have rarely been found in the involved arteries of subjects with emboli, and there has been a case report of a portion of a PFE recovered from a coronary artery. Another postulated mechanism of cardiac ischemia is prolapse of a tumor into a coronary ostia, resulting in a ball-valve phenomenon.

PFEs on the left side of the heart are felt to be symptomatic more often than right-sided lesions, with larger, mobile left-sided tumors especially vulnerable to embolization. PFEs are not felt to usually cause abnormal findings on physical examination and are not believed to usually result in significant valvular dysfunction.

Echocardiography is the diagnostic modality of choice to detect PFEs, and its increased performance and refinements in resolution are considered the main reason for the increased detection of these tumors antemortem. Transesophageal echocardiography is considered to be superior to transthoracic studies for diagnostic purposes. Although it has been claimed that there are no definitive features to distinguish PFEs echocardiographically from other cardiac masses, their appearance is characteristic enough that they usually can be identified with confidence.

The differential diagnosis of PFEs includes myxomas, vegetations, thrombi, Lambl’s excrescences, fibromas, strands, degenerative valve tissue, fenestrations, and other benign tumors. Whether or not PFEs and Lambl’s excrescences, which have been described as small fibrous tags or filiform fronds projecting from heart valves, are the same entity has been debated. Some studies have concluded that the two may be histologically similar, while other authorities consider them to be distinct entities. Lambl’s excrescences and PFEs have been described as differing in location and size, with the former usually located at the closing margins of valves and felt by some to result from turbulent blood flow and traumatic and degenerative “wear and tear.” Lambl’s excrescences are also believed to be smaller and more broader based than PFEs. Lambl’s excrescences are considered to be more common than PFEs, with prevalence estimates ranging from 14% of subjects with normal valve leaflets to 70% to 80% of adult heart
valves. As opposed to PFEs, Lambl’s excrescences are often multiple.

The embolic potential of Lambl’s excrescences seems debatable, with 1 report claiming that they embolize at a lower rate but similar fashion as PFEs, and another saying that they may not be substrates for cardiac emboli. The issue is confused further because some authorities consider PFEs to be giant Lambl’s excrescences.

Review of the literature suggests a lack of general consensus on the treatment of PFEs. Several experts agree that PFEs believed to be responsible for embolic events should be surgically excised. However, conservative treatment with anticoagulation has also been advocated with first time symptomatic lesions, with surgery recommended upon recurrent embolism. Others advocate surgical excision of all PFEs, whether symptomatic or not, due to the embolic potential.

Also advocated has been observation of asymptomatic lesions, excision of lesions 1 cm or larger, excision of all left-sided lesions, and individualizing treatment options on a case-by-case basis. It has been emphasized that even small lesions can have embolic potential. Recommendations for right-sided lesions have included excision of large, mobile symptomatic ones and observation of asymptomatic ones. Anticoagulation for PFEs has been advocated by some, either instead of or in addition to surgery.

The first reported excision of a benign heart valve tumor occurred in 1976. When excision is performed, the valve usually can be preserved, although extensive valvular involvement may necessitate replacement. There had not been reports of PFE recurrence after excision, but a recent paper describes a male who had a resection of a tricuspid valve PFE after experiencing transient ischemic attacks who was found 9 years later to have a mitral valve PFE after again presenting with neurologic embolic symptoms.

Although the natural history of benign cardiac valvular tumors is unclear, because of the potential for adverse outcomes, we must underwrite them carefully, on a case-by-case basis, until long-term studies become available that clarify this issue.

REFERENCES

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