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Cardiac Papillary Fibroelastoma: A Source of Coronary Artery Emboli and Myocardial Infarction

Gerald Fine, MD* and Shoba R. Pai, MD**

Cardiac papillary fibroelastomas, although generally small and asymptomatic, have been suspected of being responsible for cerebral accidents, angina, and sudden death. The diagnosis of tumor embolization, considered in a number of cases, has been based on circumstantial evidence in all instances except for one case of sudden death due to embolism to the left anterior

descending coronary artery. In all other cases, angina and sudden death have been related to a ball valve-type obstruction of a coronary ostium by tumor. The present case documents tumor embolism to peripheral right coronary arteries with associated, clinically silent myocardial changes.

Papillary fibroelastomas have been reported infrequently, and until their recent antemortem recognition by open-heart surgery (1) or noninvasive cardiac procedures (2-5), have been incidental findings at autopsy. Despite exposure to the stress of flow and pressure in the cardiac chambers, embolization of these tumors has been suspected infrequently and recorded only once. This report documents embolization of an aortic valve papillary fibroelastoma to the peripheral coronary circulation which resulted in myocardial damage.

Case Report

A 63-year-old, black man was hospitalized and treated in January, 1976, for pulmonary tuberculosis. Two months following cure and discharge, he returned to the emergency unit with muscular weakness, generalized fatigue, and weight loss to 98 lbs. There had been no chest pain, fever, sweats, or hemoptysis. Blood pressure was 94/70, pulse 104/min. A grade II/VI systolic ejection murmur was heard at the left sternal border.

An electrocardiogram (ECG) on May 6 and on May 12, 1976, was normal. Chest x-ray disclosed infiltrates in both lungs, more prominent in the right lower lobe. Gallium scan on July 21, 1976, revealed a triangular density in the right lower lobe which was considered to be a neoplastic process, but needle aspiration was not confirmatory. His condition deteriorated, and he expired on August 20, 1976.

Pathology

Necropsy disclosed a squamous cell carcinoma in the right lower lobe of the lung with metastases to the right tracheobronchial lymph nodes, liver, and right atrial epicardium. Bilateral organizing Klebsiella pneumonia and fibrocaseous upper lobe granulomata explained the pulmonary radiographic densities. The 375 gm heart had a thickened, left ventricular musculature with a 1 x 1.5 cm scar at the base of its posterior wall. On the right aortic cusp in the area of the

corpus Arantii was a 1.3 x 1.1 x 1 cm smooth-surfaced, glistening, bosselated, pink nodule on a short pedicle (Fig. 1). The coronary arteries, opened as far as possible, were free of emboli and showed only moderate atheromatous plaques which narrowed the lumens no more than 30%. The right coronary artery supplied the posterior wall of the left ventricle.

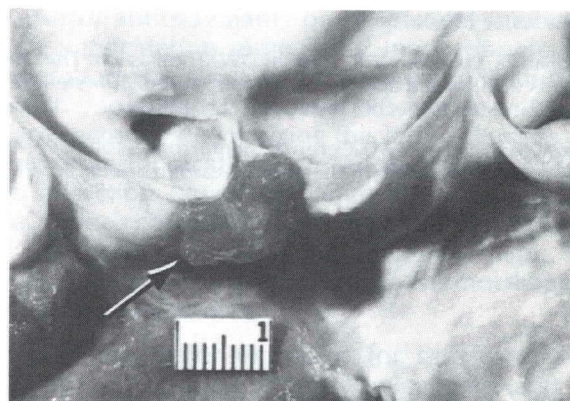


Fig. 1

Tumor nodule attached at the corpus Arantii of the right coronary cusp.

Microscopically the nodule on the aortic valve had a papillary configuration that was not discernible grossly. The cores of the papillae were dense, poorly cellular, fibrous tissue containing varying numbers of elastic fibers (Figs. 2,3). The surface of the papillary fronds were covered by single and multilayered, oval or round, endothelial-appearing cells. These cells were separated from the central connective tissue

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Cardiac Papillary Fibroelastoma

cores by varying quantities of loose, finely fibrillar or amorphous tissue that stained positively with alcian blue (pH 2.5) and colloidal iron. Prestaining incubation with 0.1% bovine testicular hyaluronidase in saline for three hours at 37°C ablated the alcian blue staining but only diminished the colloidal iron staining. The lumen of several intramuscular branches of the right coronary artery contained emboli which were histologically identical to the papillary fronds of the aortic valve tumor. A varied alteration in the adjacent myocardium accompanied the occluded arteries. Myocardial fibers were partially or completely replaced by dense or delicate fibrous tissue containing scattered hemosiderin or lipofuscin laden histiocytes and rarely polymorphonuclear leukocytes (Figs. 4,5). Emboli and myocardial changes were found in the posterior left ventricle, the free right ventricular wall, and the right atrial muscle. The sinus and atrioventricular nodes were spared. Many sections from the microscopically altered right atrial muscle had to be examined before an artery occluded with tumor was located. Emboli were not observed in the focal areas of calcification and in myocardial fibrosis that was present high in the interventricular septum or random sections of the brain and other organs.

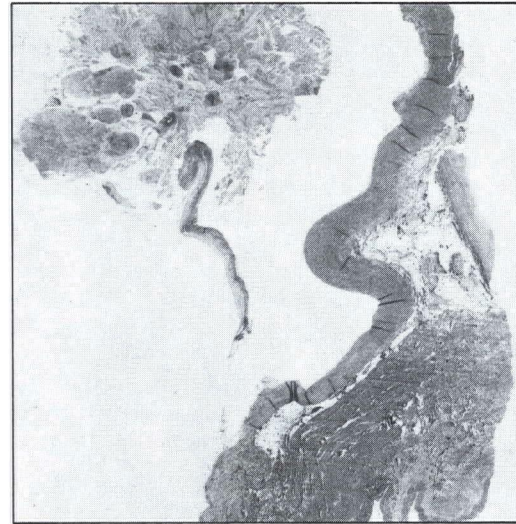


Fig. 2

The papillary character of the tumor can be seen even at this low magnification. H&E 5x.

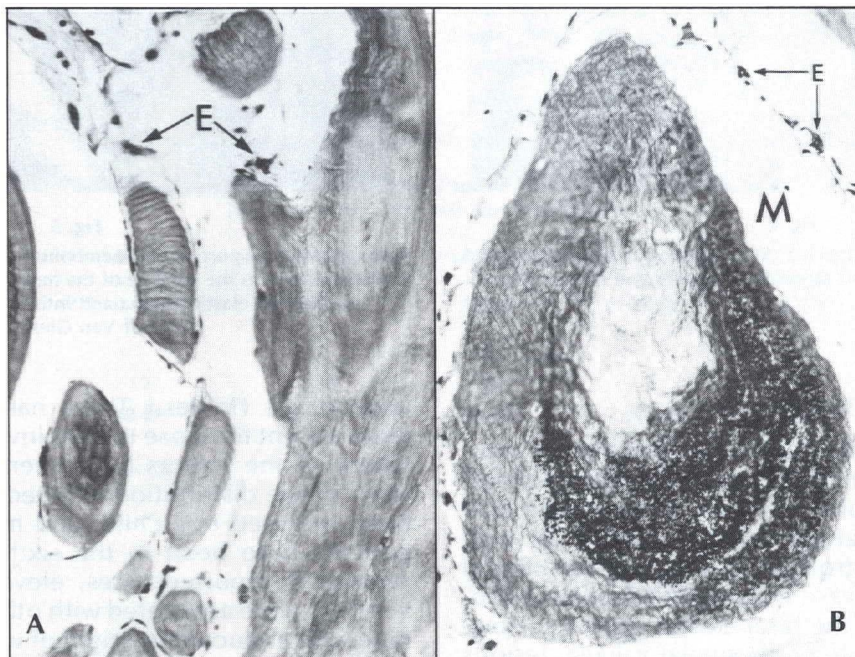


Fig. 3

A. Elastic and fibrous tissue of the poorly cellular central papillary cores are covered by endothelial cells (E). Verhoeff Van Gieson 175x.
B. Endothelial cells are separated from the core by mucin (M). 225x.

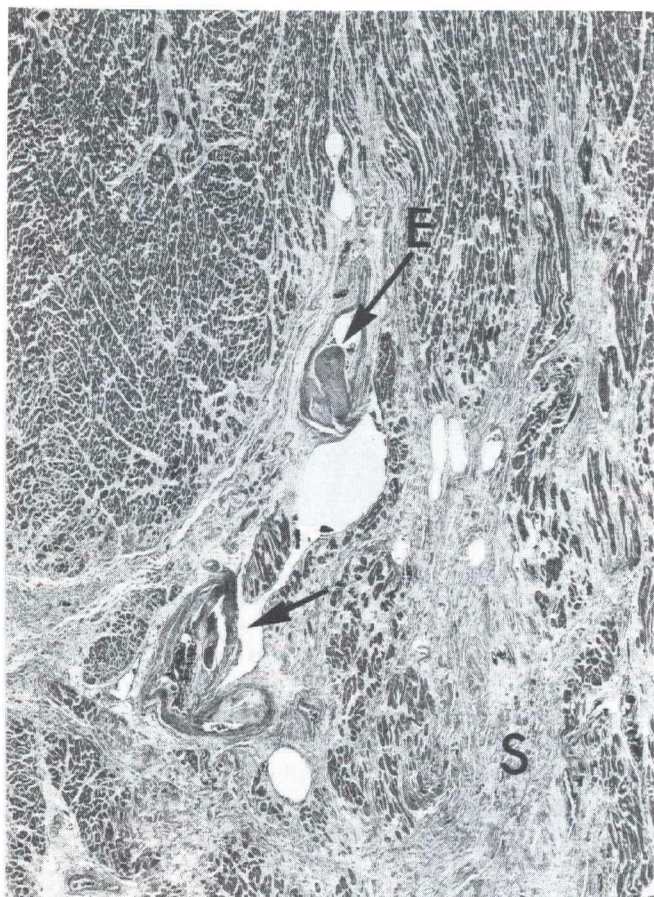


Fig. 4

Dense and delicate fibrous scarring (S) of the myocardium associated with arterial tumor emboli (arrows). Verhoeff Van Gieson 19x.

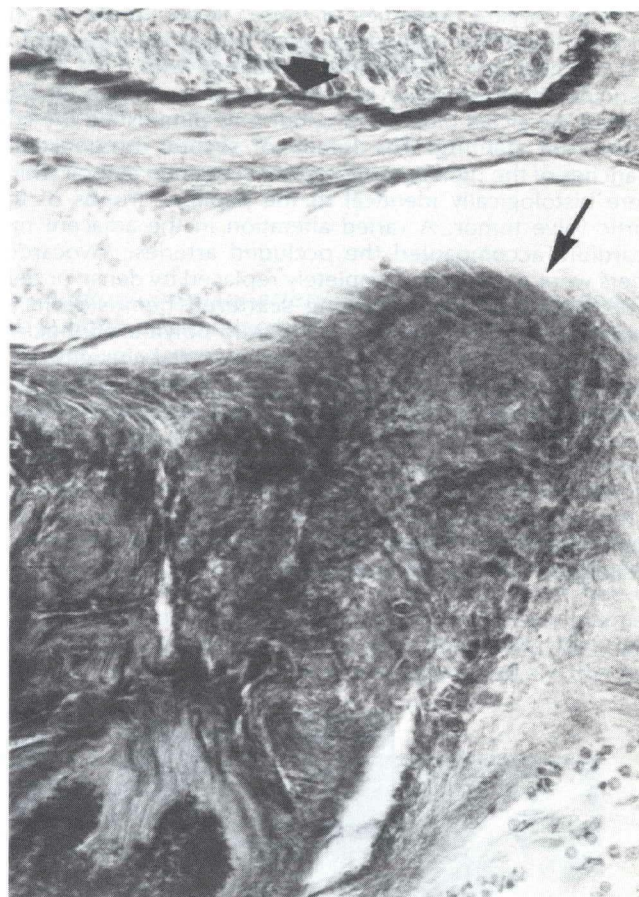


Fig. 5

Arrow illustrates a portion of the embolus E, Fig. 4, with fibrous and elastic tissue similarity to the papillae of the heart tumor (Fig. 3B). Arrowhead: internal elastic lamina and intimal fibrous thickening. Verhoeff Van Gieson 300x.

Discussion

The papillary endocardial tumor just described is morphologically distinct and differs from the polypoid myxomatous valvular nodules of infants and children (6,7) and the fibroangiomatic valvular growth (8) with which they have been grouped by some authors. Based on the infrequency of case reports, the papillary fibroelastoma ranks among the rarer cardiac growths. However, if the small, histologically similar valvular lesions found incidentally at autopsy and considered by some authors as a separate entity—Lambl's excrescence—are included, they would constitute the most frequent cardiac tumor. In the study by Dudley, et al (9), where small size was not an exclusive factor in making the diagnosis of papillary fibroelastoma, the tumor was found in 19 of 50 consecutive hearts studied. While they are more prevalent on the aortic valve, papillary endocardial tumors have been reported on all valves and in all cardiac chambers, with the exception of the atrial

appendages (Table).† There has been no sex prevalence except for those tumors involving the mitral valve where all the tumors have been found in women. A skew in age distribution is noted; only one tumor has been reported in a child, and most of the remaining patients have been in the sixth decade or beyond. Among 56 reported cases, eleven tumors in the left ventricle were associated with other cardiac pathology: nine with endocarditis [eight of which were designated as rheumatic (3,10,11)] and one each with prolapsed mitral valve (5) and hypertrophied ventricular septum (4). One right ventricular tumor was accompanied by an interventricular septal defect (1).

Papillary fibroelastomas have been considered to be responsible for various clinical and morphologic alterations, but with the exception of one case, evidence of

†A tumor in the left atrium has been recorded in the Armed Forces Institute of Pathology collection (17).

Cardiac Papillary Fibroelastoma

tumor embolization has been circumstantial. Nassar and Parker (10) suspected cerebral embolization in three cases, but the brain was not examined in two cases. One tumor involved the aortic valve, and another involved the tricuspid valve, a site unlikely to be responsible for cerebral embolism in the absence of a communication between right and left cardiac chambers. In the third case, there was involvement of the papillary muscle of the left ventricle, but no emboli were found in the focal encephalomalacia.

Among seventeen papillary tumors of the aortic valve in 56 reported cases (Table), there was associated angina and/or sudden death in six instances. Angina in four patients (5,12,13,14) was the result of an apparent intermittent ball valve occlusion of a coronary ostium. In one of these, the angina was associated with a surgically removed 1.5 x 1 cm sessile tumor which was attached to the noncoronary cusp and, before excision, could not be manipulated to occlude either coronary ostium (5). However, the patient was free of symptoms in the week following removal of the tumor.

Sudden death was attributed to tumors in four cases. In one case, a tumor from the right coronary cusp filled the proximal portion of the left anterior descending coronary artery (15), and in two instances a ball valve

occlusion of the right coronary ostium appeared to be the cause for the patient's demise (12,13). In the fourth case there was tumor on the left posterior aortic cusp and occlusion of the right coronary ostium, but it was not clear whether the occlusion was due to the tumor or to arteriosclerosis (18).

In the present case no clinical symptoms were referable to the heart, and ECGs were normal on two occasions, one week apart, three months before the patient's death. Some of the histologic myocardial changes suggest that emboli may have been present at the time of the ECGs. With the exception of the changes high in the interventricular septum, tumor emboli were found in all areas where there was microscopic myocardial damage. Similar involvement of the myocardium was found in areas supplied by the right coronary artery whose ostium was in close proximity to the tumor. These findings, together with the minimal evidence of atheromatous change in the coronary arteries, strongly suggest a causal relationship between the emboli and the myocardial alterations. Subserial sectioning of the altered right atrial myocardium to uncover the arterial embolus in this area indicates that an exhaustive microscopic search may be required to locate the arterial emboli of papillary fibroelastoma.

TABLE
Fifty-six Reported Cardiac Papillary Fibroelastomas
Size, Anatomic, Sex, and Age Distribution

	Aortic Valve (2,5,10,12-14,15)	Mitral Valve (5,10)	Pulmonary Valve (2,10)	Right Heart* (1,10,16)	Left Ventricle (2-4,11)	Tricuspid Valve (2,10,11)
Men						
Number	12	0	8	2	2	3
Age (yrs)	38-89		53-82	52,72	46,64	83**
Size of Tumor (cm)	0.4-1.5		0.1-1.4	0.7,1.2	0.7,1.5	0.3-1.3
Women						
Number	5	7	5	2	6	4
Age (yrs)	45-67	9 mos-83	19-68	80,92	29-71	35-62
Size of Tumor (cm)	0.5-0.8	0.5-1	0.5-1	0.1,3.5	1-4	0.6**

*Two tumors in right atrium; two in right ventricle.

**One age or measurement recorded.

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