

Embolic Stroke From Cardiac Papillary Fibroelastomas

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We describe two patients with cerebral emboli originating from a cardiac papillary fibroelastoma and compared them with six patients reported in the literature. Surgical excision was curative in both of our patients. The surface topography of the tumor, as visualized by scanning electron microscopy, is described. (*Stroke* 1988;19:1171–1173)

Cardiac papillary fibroelastomas are rarely encountered and, before 1981, were diagnosed only at autopsy.¹ The lesion consists of a frond-like papillary growth, which typically occurs on the cardiac valves but can also appear over the papillary muscles, chordae tendineae, ventricular septum, or the endocardial surface.² Although it has been recognized that occlusion of the coronary ostia by papillary fibroelastomas may rarely cause sudden death,³ the neurologic complications of this lesion have not been widely appreciated. We describe two patients in whom emboli originating from papillary fibroelastoma caused transient ischemic attacks (TIAs) and cerebral infarction.

Case Reports

Case 1. A 43-year-old Caucasian woman was found paretic and unresponsive by her daughter. Her neurologic deficits had totally resolved by the time of admission to the emergency department 4 hours later. Adult-onset diabetes mellitus was the only recognized risk factor for cerebrovascular disease. In addition to insulin, the patient had been taking 50 mg dipyridamole t.i.d. for several years. The examination was remarkable for obesity. No abnormalities were detected on cardiac examination, and there were no carotid bruits. The neurologic examination was normal. The following diagnostic studies were within normal limits: computed tomogram (CT scan) of the head, platelet count, electrocardiogram, electroencephalogram, oculoplethysmography, and 24-hour Holter monitoring. Lipid profile demonstrated slightly elevated high density lipoprotein. An echocardiogram

revealed an echogenic mass measuring 2 × 3 cm on the anterior leaflet of the mitral valve near the lateral commissure. The intracardiac tumor was resected on July 5, 1985, with the complication of a transient right hemiparesis and brief focal seizure. Several follow-up CT scans remained normal. Gross pathologic exami-

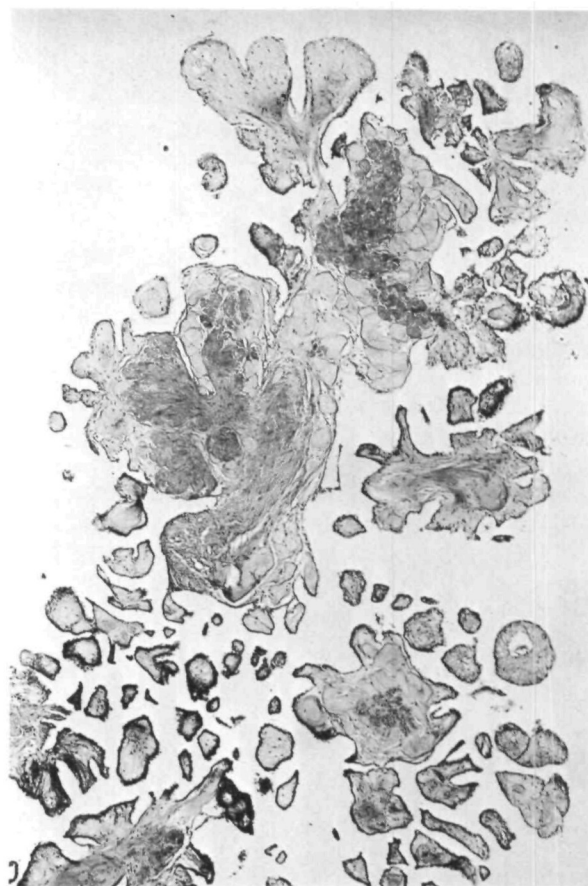


FIGURE 1. Case 1. Light micrograph of cross-sectioned tumor showing variably thick avascular core covered by single layer of endothelial cells. Periodic acid-Schiff stain, ×17.5.

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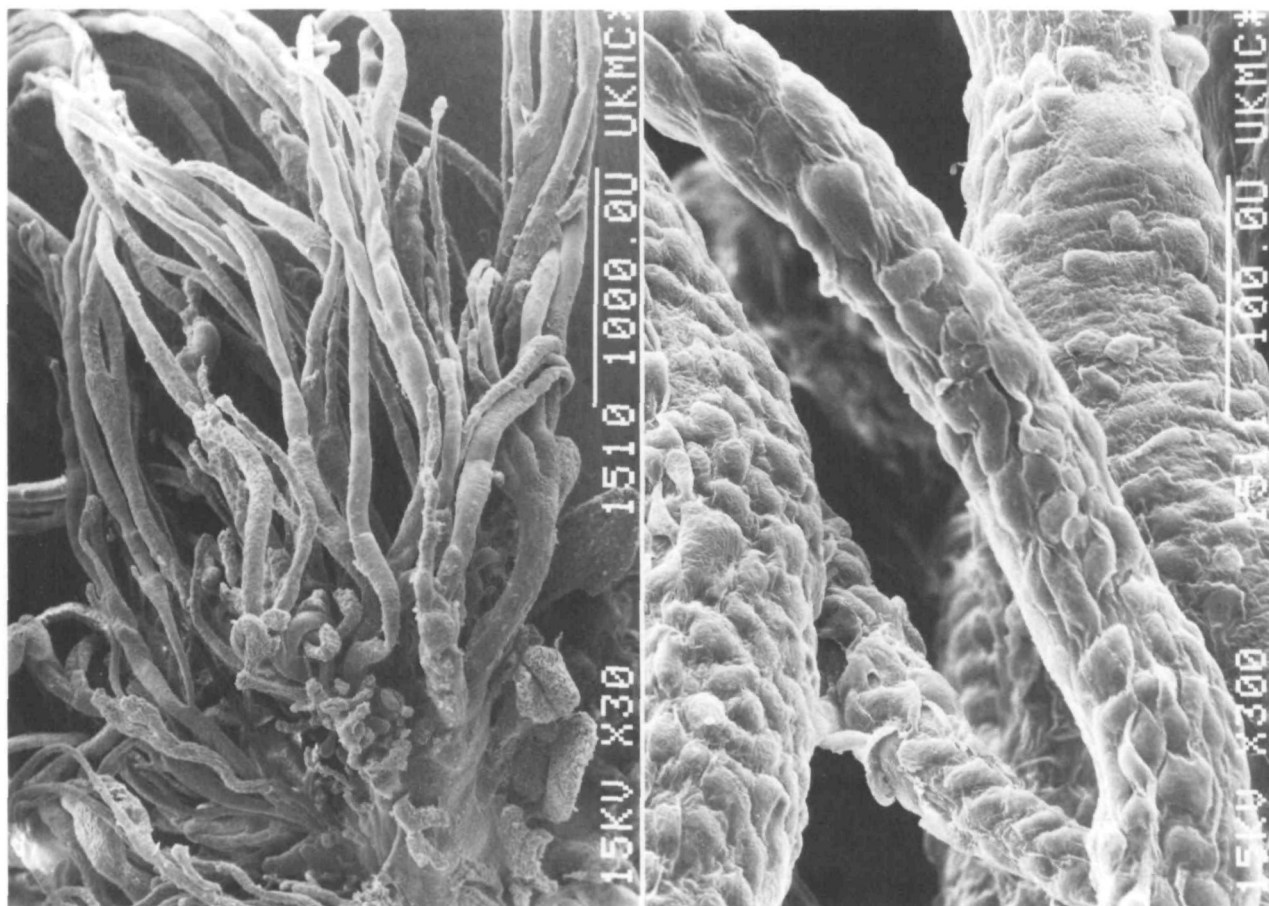


FIGURE 2. Case 1. Left: Low-power scanning electron micrograph showing elongated, tapered, and sometimes bunching fronds radiating from short central base. $\times 21$. Right: High-power scanning electron micrograph of individual hair-like projections showing plump, paved covering of endothelial cells. $\times 213$.

nation of the tumor revealed a mass, 1.8 cm in diameter, on a stalk with tiny hair-like appendages producing a sea anemone configuration. Histologic examination showed a typical papillary fibroelastoma consisting of nonvascularized cores of fibroelastic tissue covered with a loose myxoid layer and endocardial lining cells² (Figure 1). Scanning electron microscopy revealed variations in the thickness of the papillae, with some bunching and club-shaped tips partly covered by endocardial cells (Figure 2).

Case 2. This 52-year-old Caucasian man has a history of multiple cerebral infarcts occurring in

July of 1981, November of 1982, and February of 1983. On one occasion, the infarction was complicated by tonic/clonic seizures requiring phenytoin and phenobarbital. He was admitted in August of 1984 after experiencing a TIA referable to the vertebrobasilar vascular system, manifested by tetraparesis without loss of consciousness lasting approximately 1 hour, followed by complete resolution of the new deficits. CT scans of his head with and without contrast (Figure 3) revealed five low-density, nonenhancing lesions involving the entire right occipital cortex, the anterior left occipital

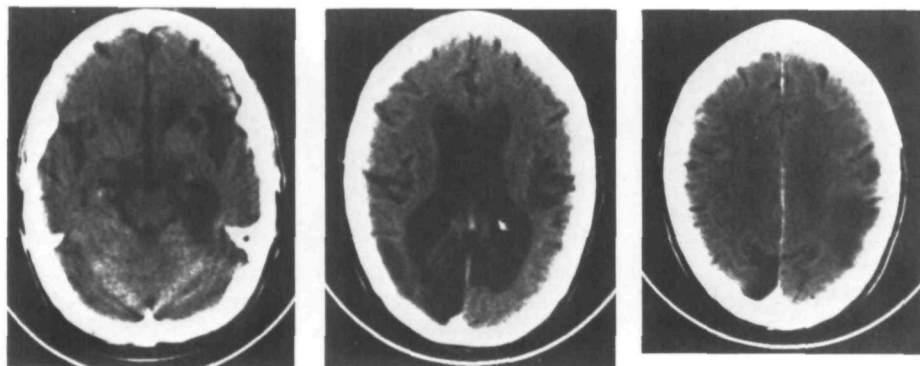


FIGURE 3. Case 2. Computed tomogram of head demonstrating multiple infarctions in occipital cortex bilaterally, in left thalamus, in right insula, and in left parietal lobe.

TABLE 1. Reports of Papillary Fibroelastoma and Stroke

Author	Year	Patient's age	Patient's sex	Previous TIA or stroke	Infarct distribution
Yater ⁴	1931	70	M	7	LMCA
Abu Nassar and Parker ⁵	1971	54	M	?	LMCA
Fowles et al ⁶	1981	35	M	Yes	LMCA
Ong et al ⁸	1982	71	F	Yes	RMCA
Topol et al ⁹	1986	27	M	No	LMCA
McFadden and Lacy ¹⁰	1987	41	F	No	LMCA
This report	1988	43	F	No	TIA only
		52	M	Yes	Basilar, L MCA, R MCA

TIA, transient ischemic attack; M, male; F, female; L, left; R, right; MCA, middle cerebral artery.

cortex, the left thalamus, the right insula, and the left parietal lobe. A four-vessel cerebral angiogram established that the carotid and vertebrobasilar arterial systems were normal. An echocardiogram delineated a 1.5 x 2 cm mass on the posterior mitral valve that was removed on August 31, 1984, without complication. Pathologic examination revealed a frond-like tumor 1.3 cm in diameter on a stalk that on microscopic examination had an arborescent core and was consistent with a papillary fibroelastoma. No further cerebrovascular events have occurred during the 3-year postoperative follow-up.

Discussion

The papillary fibroelastoma is generally small (0.1-4.0 cm in diameter) and accounts for only 1% of primary heart tumors. It arises from the valvular connective tissue and is anchored by a short pedicle.⁴ Multiple hair-like fronds radiate from an avascular central fibrocollagenous core (Figures 1 and 2, left), and these villous structures are invested by a single layer of endocardial cells (Figure 2, right).⁵ Fibrin thrombi may adhere to some of the papillary structures,⁶ which presumably are the source of emboli.

Traditionally, papillary fibroelastomas have been considered to be benign lesions, discovered as incidental findings at autopsy. As recently as 1982, the tumor was visualized before surgery in only three instances.⁷ Although their potential for causing sudden death by occluding a coronary ostium has been recognized,³ the occurrence of stroke from emboli originating from this tumor has been infrequently documented. Table 1 summarizes the six cases with papillary fibroelastoma and TIA/stroke reported in the literature compared with the two cases we describe. Although there is a male preponderance in this group (five men, three women), no obvious sex predilection has been recognized in larger autopsy series. The average age of the patients was 49 (range 27-71) years. Fifty percent of the patients had TIAs or previous strokes prior to discovery of the papillary fibroelastoma. In our Case 2, the causal relation between the mitral papillary fibroelastoma and the multiple infarcts is

particularly clear based on the history of three previous vascular events, the CT visualization of multiple old cerebral infarctions in several vascular territories (Figure 3), the normal extracranial vessels, and the absence of further vascular events during a 3-year follow-up after the tumor was removed. Thus, it appears that papillary fibroelastomas carry a definite, albeit low, potential for cerebral embolization and should be included in the list of cardiac tumors causing stroke.

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KEY WORDS • embolization • endocardial fibroelastosis • cerebral infarction • transient ischemic attack