Commentary: Benign cardiac papillary fibroelastomas: Beware the “divergents”

David D. Yuh, MD, FACS, FACC

From the Department of Surgery, Stamford Hospital, Stamford, Conn.

Disclosures: Author has nothing to disclose with regard to commercial support.

Received for publication Oct 20, 2018; accepted for publication Oct 21, 2018; available ahead of print Nov 30, 2018.

Address for reprints: David D. Yuh, MD, FACS, FACC, Department of Surgery, Stamford Hospital, Stamford, CT

(E-mail: DYuh@stamhealth.org).

J Thorac Cardiovasc Surg 2019;157:e129-30

0022-5223/\$36.00

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<https://doi.org/10.1016/j.jtcvs.2018.10.098>

David D. Yuh, MD, FACS, FACC

Central Message

Cardiac papillary fibroelastomas can be multicentric, with both local and distant recurrence, warranting better insight into their bioepidemiologic profile and improved resection techniques.

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The rarity, indolent progression, and “benign” classification of most primary cardiac tumors has likely contributed to the lack of understanding of their histogenesis, biologic behavior, and treatment relative to other more prevalent noncardiac tumors. Because they are viewed more as clinical curiosities than as bona fide cardiac pathologies, with the primary treatment consisting of surgical excision, the literature concerning papillary fibroelastomas has largely consisted of clinical series and case reports of unusual locations and manifestations,^{1,2} such as the report in this issue of the *Journal* by Popovic and colleagues.³ What distinguishes this case report from others, however, is its description of recurrence of a valve-based (aortic valve) papillary fibroelastoma to a “distant” nonvalvular site (left ventricle). This finding is inconsistent with the notions that these lesions are benign and may not even be true neoplasms but rather hamartomas or reactive changes of normal tissue. Importantly, it implies that, like most other neoplasms, the biologic behavior and metastatic potential of papillary fibroelastomas may actually vary on a spectrum.

Playing devil’s advocate, one cannot help but speculate that, rather than a distant recurrence, this was an atypical asynchronous or metasynchronous multifocal presentation of papillary fibroelastoma, with microscopic foci of fibroelastoma cells in the left ventricle that could not be visualized during the primary resection of the valvular lesions but later developed into the impressive tumors nicely depicted in this report. Although multiple tumors are uncommon with papillary fibroelastoma,⁴ an incidence as great as 6% was noted in one large study.⁵ Certainly, multicentricity is also observed with myxomas, another benign cardiac tumor, and is associated with a more aggressive bioepidemiologic profile, including familial patterns of inheritance (5%-10% of cases), higher recurrence rates (12%-22%), and presentation at a younger age.⁶

It is tempting to postulate that multiple lesions on initial presentation, as observed here, may predispose to local or distant recurrence. It would be interesting to determine

whether reported cases of recurrence were associated with multiple tumors at primary resection, as at least 1 of them was.⁷ Multiple tumors may be a manifestation of more aggressive proliferative behavior and may warrant more frequent postoperative echocardiographic surveillance. The association of multicentric tumors on initial presentation as a risk factor for recurrence has been reported with other neoplasms, including breast and papillary thyroid carcinoma.^{8,9}

Because incomplete resection has been implicated in local recurrence of both cardiac myxomas and papillary fibroelastomas,¹⁰ additional consideration to the surgical technique of resection, including valve-preserving methods and intraoperative frozen-section analysis to ensure complete resection, may be warranted. This case report suggests, however, that complete surgical extirpation of the primary tumor may still be inadequate in preventing distant recurrence, because the affected aortic valve was completely excised.

Although the risk of recurrence for the most common benign cardiac tumors remains very low, this case report illustrates that the morbidity of local or distant recurrence is significant and warrants more incisive investigation into their histopathologic behaviors and genomics to identify more aggressive subsets. Moreover, a closer look at developing techniques for ensuring complete resection of these tumors may further reduce recurrence rates.

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