

Aortic Valve Papillary Fibroelastoma

A Rare Cause of Angina

Vito D. Bruno, MD
Giovanni Mariscalco, MD, PhD
Stefano De Vita, MD
Gabriele Piffaretti, MD
Daniele Nassiacos, MD
Andrea Sala, MD

A 66-year-old woman with diabetes mellitus and a history of hypertension presented with intermittent episodes of chest pain and palpitations. An electrocardiogram showed no previous ischemic events but revealed atrial fibrillation. Transthoracic echocardiography disclosed a round, mobile mass on the aortic valve. Transesophageal echocardiography showed the mass (14 × 10 mm) on the aortic side of the right coronary cusp (Fig. 1). It was unclear whether the mass was a thrombus, a tumor, or a vegetation, but given its position and mobility, urgent surgical excision was planned.

The mass was connected to the free margin of the right aortic valve leaflet and threatened to obstruct the right coronary ostium (Fig. 2). A complete excision was performed, and the valve leaflets were spared. Macroscopically, the mass resembled a sea anemone (Fig. 3), which supported a diagnosis of papillary fibroelastoma (PFE). Histopathologic evaluation confirmed the diagnosis.

Comment

Papillary fibroelastoma is the 3rd most common primary cardiac tumor¹ and the most frequent valvular tumor.² Clinical presentation varies widely, from asymptomatic to severe embolic sequelae. Coronary occlusion by an aortic PFE has rarely been reported.

Section Editor:

Raymond F. Stainback, MD,
Department of Adult
Cardiology, Texas Heart
Institute at St. Luke's
Episcopal Hospital, 6624
Fannin St., Suite 2480,
Houston, TX 77030

From: Department of Surgical Sciences (Drs. Bruno, Mariscalco, Piffaretti, and Sala), University of Insubria Varese University Hospital, 21100 Varese; and Department of Cardiology (Drs. De Vita and Nassiacos), Presidio Ospedaliero di Saronno, 21047 Saronno; Italy

Address for reprints:

Vito Domenico Bruno, MD,
Department of Cardiac
Surgery, Varese University
Hospital, via Giucchiardini 7,
21100 Varese, Italy

E-mail: vitodomenicobruno@gmail.com

© 2011 by the Texas Heart®
Institute, Houston

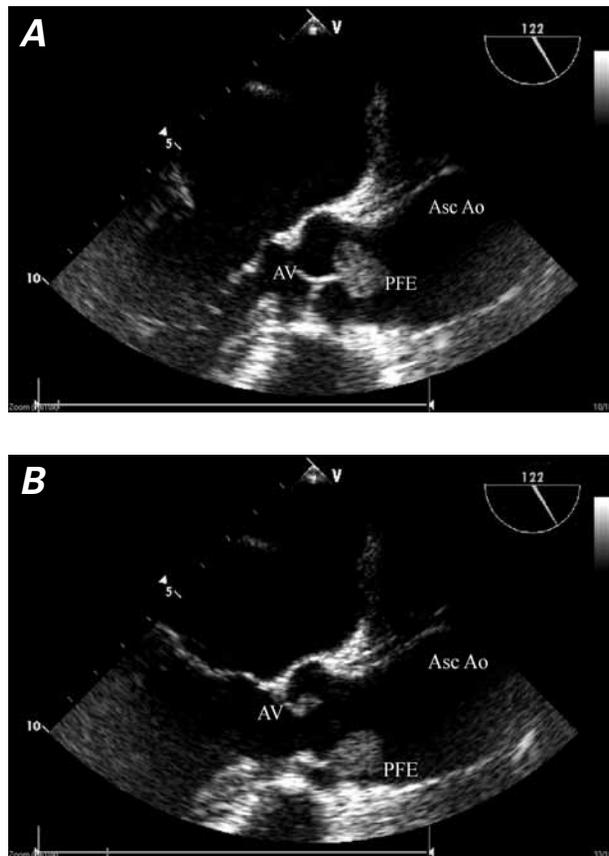


Fig. 1 Preoperative transesophageal echocardiograms (mid-esophageal long-axis views) show the mass on the aortic valve during **A**) diastole and **B**) systole.

Asc Ao = ascending aorta;
AV = aortic valve; PFE =
papillary fibroelastoma

Real-time motion image is
available at www.texasheart.org/journal.

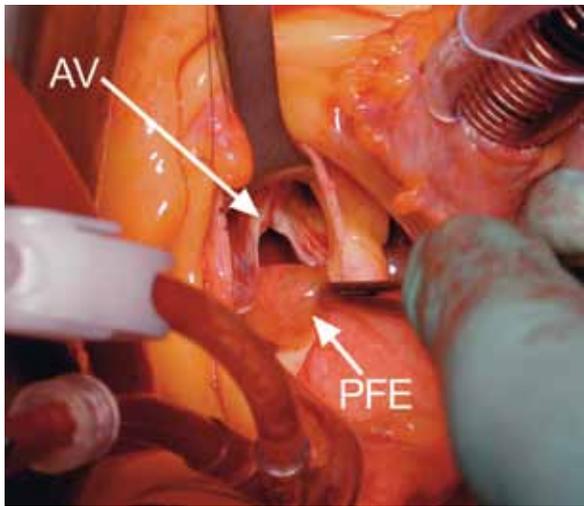


Fig. 2 Intraoperative photograph shows a round, mobile mass on the free margin of the right coronary cusp of the aortic valve. AV = aortic valve; PFE = papillary fibroelastoma



Fig. 3 Macroscopic appearance of the neoplasm, in saline solution. The multiple frond-like structures give the appearance of a sea anemone.

In 1984, it was reported that an otherwise healthy man suddenly died when a PFE occluded the left coronary ostium.³ A similar case was described in 1985.⁴ Nonfatal sequelae such as dynamic coronary obstruction, anginal attacks, and recurrent myocardial infarction have also been reported. However, cerebral embolization is the most frequent sequela of PFEs. Surgical treatment is typically curative and yields good long-term results.

References

1. Grinda JM, Couetil JP, Chauvad S, D'Attellis N, Berrebi A, Fabiani JN, et al. Cardiac valve papillary fibroelastoma: surgical excision for revealed or potential embolization. *J Thorac Cardiovasc Surg* 1999;117(1):106-10.
2. Mariscalco G, Bruno VD, Borsani P, Dominici C, Sala A. Papillary fibroelastoma: insight to a primary cardiac valve tumor. *J Card Surg* 2010;25(2):198-205.
3. Seib HJ, Wildenauer M, Luther M, Pitzl H, Henselmann L. Papillary fibroelastoma of the aortic valve. Sudden death caused by an uncommon tumor of the heart [in German]. *Z Kardiol* 1984;73(6):409-13.
4. Zull DN, Diamond M, Beringer D. Angina and sudden death resulting from papillary fibroelastoma of the aortic valve. *Ann Emerg Med* 1985;14(5):470-3.