Hook-Like Long Linear Echogenic Mobile Mass Arising in the Left Ventricular Outflow Tract Presenting With Orthostatic Hypotension and Syncope – Diagnosis on Multimodal Imaging Echocardiography and Computed Tomography –

Eun Jeong Cho, MD; Sung-Ji Park, MD; Mi-Rae Lee, MD; Pyo Won Park, MD; Seung Woo Park, MD; Yeon Hyeon Choe, MD; Su-Jin Kim, RN; YuRim Han, RN

Figure 1. (A, B) Transthoracic echocardiography and (C, D) 3-D echocardiography showing a mobile, linear pedunculated mass on (A) parasternal long axis; (B, D) 5-chamber view; and (C) parasternal short axis. (E) Multiplane trans-esophageal echocardiogram showing a linear tumor attached to the lateral aspect of the left ventricular wall, in the left coronary cusp area (systolic phase, at an angle of 135°); (F) diastolic phase, at an angle of 135°. (G, H) 3-D echocardiogram, at an angle of (G) 120° and (H) 135°.

Cardiac papillary fibroelastomas (PFEs) are rare benign tumors arising primarily from the valvular endocardium. In contrast, non-valvular PFEs – that is, PFEs attached to the left ventricular outflow tract (LVOT) – are rare. Most PFEs have been discovered incidentally, but rarely they are accompanied by seemingly unrelated, atypical symptoms of orthostatic hypotension or syncope. A diagnosis of PFE is typically reached via transthoracic echocardiography.
Figure 2. Contrast-enhanced electrocardiography-gated multislice spiral computed tomography: (A) sagittal, left ventricular (LV) outflow tract level; (B) 3-dimensional image. (C) Intraoperative view of the aortic valve, showing a linear mass attached to the lateral aspect of the LV wall, in the left coronary cusp area. (D,E) Histological section of the excised mass showing papillary formation with homogenized stroma and slightly broadened endothelial cells at the surface (H&E). Original magnification (D) ×40, (E) ×200.

Additional imaging was then performed, including multi-plane TEE (Figures 1E,F) and 3-D echocardiography of TEE (Figures 1G,H), which better demonstrated that this mass was attached to the lateral aspect of the LV wall. Regurgitation through the aortic valve was also noted during the systolic phase. The hook-like tumor was attached to the lateral aspect of the LV wall. A common bundle, called the bundle of His, was located at the basal ventricle, and propagation from the AV node to the ventricles was provided by the common bundle, therefore it was thought that the hook-like tumor disturbed propagation from the AV node to the ventricles, causing possible bradycardia or arrhythmia. This could generate the symptoms of syncope or orthostatic hypotension.

Contrast-enhanced ECG-gated 64-slice spiral CT showed a well-defined, 14–17-mm hook-like long linear mass attached to the lateral aspect of the LV wall, in the left coronary cusp area (Figures 2A,B). Because of the associated symptoms, surgical resection was performed under cardiopulmonary bypass. Intraoperatively, a 45×2-mm-long linear mass was found attached to the lateral aspect of the LV wall, in the left coronary cusp area. On palpation, the mass was soft and slippery and had an appearance similar to a long string (Figure 2C). After the mass was completely resected, histopathology confirmed the diagnosis as a benign PFE, specifically showing a central avascular collagenous core of dense connective tissue surrounded by a peripheral layer of loose connective tissue (Figures 2D,E). The patient was then discharged uneventfully 8 days later. At 3-month follow-up, ECG showed normal sinus rhythm, and no evidence of sinus pause or second-degree AV block was observed on
Hook-Like Mass in LVOT With Hypotension, Syncope

24-h ECG Holter monitor.

Although PFEs are generally benign and lack any specific clinical features, early diagnosis is important because these tumors represent a surgically correctable cause of systemic emboli, stroke, myocardial infarction, and sudden cardiac death. Primary surgical excision is recommended for symptomatic PFEs.

PFEs represent rare cardiac tumors that mainly evolve in non-valve areas. These tumors are usually discovered incidentally due to their small size and tend to be attached to the mobile valve structure. Rarely, however, they are accompanied by atypical symptoms of orthostatic hypotension, syncope and so on. Although PFEs are relatively benign and lack specific clinical features, critical complications have been associated with these neoplasms. Even when an atypical symptom is present, appropriate use of imaging modalities and urgent surgical treatment can reduce the severity of complications and save lives.

References