Cardiac papillary fibroelastoma (PFE) is a rare benign tumor that originates mainly from the aortic valve, especially the aortic and mitral leaflets. PFEs are usually not observed on computed tomography (CT) or magnetic resonance imaging (MRI) because they are small and attached to moving valves. No clinical feature is specifically suggestive of PFE, and these tumors are usually found incidentally at the time of thoracic imaging using CT, MRI, and/or echocardiography. PFEs arising from the aortic valve, however, have been implicated in the occurrence of sudden death by causing...
obstruction of the ostium of the right or left coronary arteries.\textsuperscript{3,4} In this case report, we describe a very large, popcorn-like PFE that was diagnosed using a multimodal imaging strategy, and discuss the detection and management of this disease.

A 53-year-old man who underwent right upper lobectomy due to adenocarcinoma of the lung was referred for further evaluation of a mass-like lesion in the aortic valve that was detected on CT (Figure 1A). The patient’s primary complaint was mild dyspnea on effort. He denied neurological symptoms and his level of activity was normal. Physical examina-
tion and laboratory findings were unremarkable. Furthermore, electrocardiography (ECG) and chest radiography results were normal.

Transthoracic echocardiography demonstrated a huge, floating mobile mass (20×15 mm) attached to the aortic valve by a stalk (Figures 1B,C). A popcorn ball-like mass attached to the right coronary sinus with a central necrotic area was seen more clearly on multi-plane transesophageal echocardiography (Figures 1D,E) and on 3-D echocardiography (Figure 1F). Except for the non-coronary cusp, the aortic valve and other cardiac structures were normal. There was no regurgitation through the aortic valve.

Contrast-enhanced ECG-gated 64-slice spiral CT showed a well-defined, pedunculated and spherical lesion in the commissure of the right coronary and non-coronary aortic valve leaflets (Figures 2A,B). The lesion, which had a relatively homogenous inner structure and a slightly villous outer margin, had a density of 60 HU. To further characterize and diagnose differentially with metastasis of the mass, ECG-gated MRI was performed. This showed a solid mass with intermediate signal intensity on T1- and T2-weighted spin echo imaging (Figures 2C,D).

Surgical resection under cardiopulmonary bypass was performed. On exploration, a 20×13-mm pedunculated mass attached to the commissure between the non-coronary cusp and right coronary cusp was found (Figure 2E). The mass was soft, slippery, and had numerous filaments. Complete resection of the mass, including the stalk, was achieved. Histopathology of the tumor confirmed that it was a benign PFE comprising a central avascular collagenous core of dense connective tissue surrounded by a peripheral layer of loose connective tissue (Figure 2F). On Elastica van Giesen stained section, the papillary surface was covered by a layer of endothelial cells and had fibrous stroma, including elastic fibers (Figures 2G,H). The patient was discharged uneventfully 7 days after resection.

Cardiac PFE is a rare benign tumor arising primarily from the valvular endocardium, especially the aortic valve or mitral valve.1 Most PFEs are discovered incidentally during evaluation of unrelated medical problems due to their small size and tendency to be attached to mobile valves. Although PFEs are benign, they can cause serious complications such as thromboembolism, myocardial ischemia, stroke, and sudden death.6,7 PFEs are usually diagnosed on transthoracic or transesophageal echocardiography. Furthermore, 3-D echocardiography can help to clarify the relationship between the surrounding tissues and the tumor. Recently, multi-slice spiral CT and MRI have also been used to better delineate similar tumors and to obtain more precise anatomical information.7 Although PFEs are often readily identified by echocardiography alone, use of multiple imaging modalities such as 3-D echo, MDCT, and MRI can unambiguously distinguish these lesions from other cardiac lesions.6,8

Surgical excision is recommended for symptomatic PFEs.9 For asymptomatic patients, however, the treatment course is less obvious. Considering the friable nature, large size, and mobility of the tumor in the present case, it was surgically resected to prevent potential complications such as embolization and sudden cardiac death. A simple shave excision technique with a margin of normal endocardium is the most common technique used to surgically excise PFEs.1

To summarize, we characterized a tumor identified incidentally on routine follow-up chest CT using a multimodal imaging approach, which allowed identification of the mass as a PFE. The case described here is rare in terms of the large size and unusual shape of the PFE.

References