

## Coexistence Case of Papillary Fibroelastoma and Lambl's Excrescence of the Left Heart, Which Is Occasionally Associated with Significant Stroke

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### Summary

Papillary fibroelastoma (PFE) is a benign tumor that arises mostly from left-sided valves. PFE can cause stroke, and surgical resection may be needed. Lambl's excrescence (LE) is a filiform valvular lesion and is considered a possible cause of stroke. A 79-year-old man with light-headedness and left-sided hemiparesis was diagnosed with stroke. Transesophageal echocardiography (TEE) revealed a round-shaped mobile mass in the left ventricular outflow tract (LVOT), which was considered the cause of the stroke. Surgical resection was performed transaortically, and during surgery, a mass was incidentally detected on the noncoronary cusp (NCC), which was also resected followed by aortic valve replacement. Pathology confirmed that the mass in the LVOT was a PFE and that the filiform mass on the NCC was LE. We herein report a rare case of PFE in the LVOT and coexisting LE on the NCC. A careful examination via TEE helps to identify other possible causes of stroke hidden behind the obvious cause.

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**Key words:** Clinical pathology, Transesophageal echocardiography

Most cardiac tumors are metastatic cancers, and primary cardiac tumors are rare entities with an incidence of 1.38 to 30 out of 100,000 people per year. The 3 most common primary tumors in people aged 16 and older are myxomas (50%), lipomatous tumors (21%) and papillary fibroelastomas (16%).<sup>1</sup> Papillary fibroelastoma (PFE) is a benign cardiac tumor that often arises on the aortic and mitral valves. Gowda, *et al.* showed that among 611 patients, 511 patients had tumors on valves (84%), 223 had tumors on aortic valves (44%), and 180 had tumors on mitral valves (35%). For nonvalvular PFE, the left ventricular wall was the most common site (55 patients).<sup>2</sup> Lambl's excrescence (LE) is a filiform valvular frond that is similar to PFE and can cause embolism.<sup>3</sup> We report the case of simultaneous removal of PFE found at a rare site on the left ventricular outflow tract (LVOT) and of LE found incidentally during surgery. A careful inspection should be performed because PFE can cause us to overlook other significant causes of embolism.

### Case Report

A 79-year-old male presented with light-headedness

that had started 5 hours before arriving at our hospital. His past medical history included emphysema, hypertension, dyslipidemia and hyperuricemia, and he is currently taking vilanterol, fluticasone, theophylline, nifedipine, and pravastatin but has no history of stroke. No relevant family history was provided other than his 60 years of smoking. His vital signs on admission were unremarkable except for a high blood pressure of 173/87 mmHg. A slightly altered mental status was observed (Glasgow Coma Scale of 14). Neurological examination indicated left hemiparesis and left unilateral spatial neglect. Laboratory data revealed that his hemoglobin A1c level was 6.3%, his low-density lipoprotein cholesterol level was 130 mg/dL, his B-type natriuretic peptide level was 33.8 pg/mL, and his D-dimer level was 0.70 µg/mL. Antinuclear antibody tests were negative, and his blood cultures were sterile. Magnetic resonance imaging (MRI) revealed an infarct in the right corona radiata and parietal lobe (Figure 1A, B). Magnetic resonance angiography revealed right middle cerebral artery occlusion (Figure 1C), and the diameter of the right middle cerebral artery was approximately 3 mm. Electrocardiography showed sinus rhythm and complete right bundle branch block. Chest radiogra-

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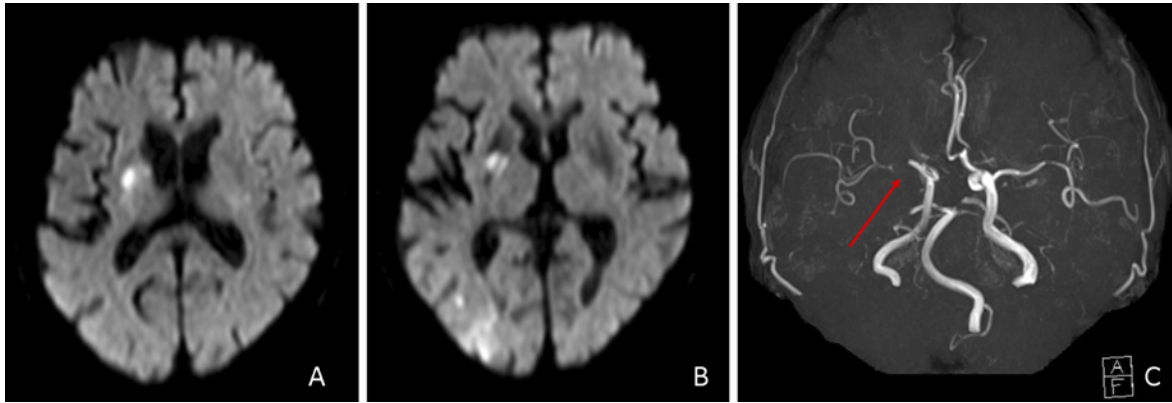
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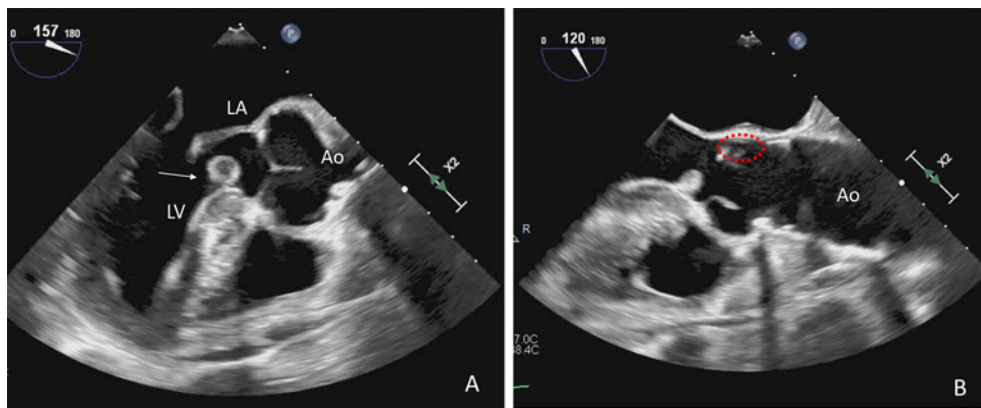
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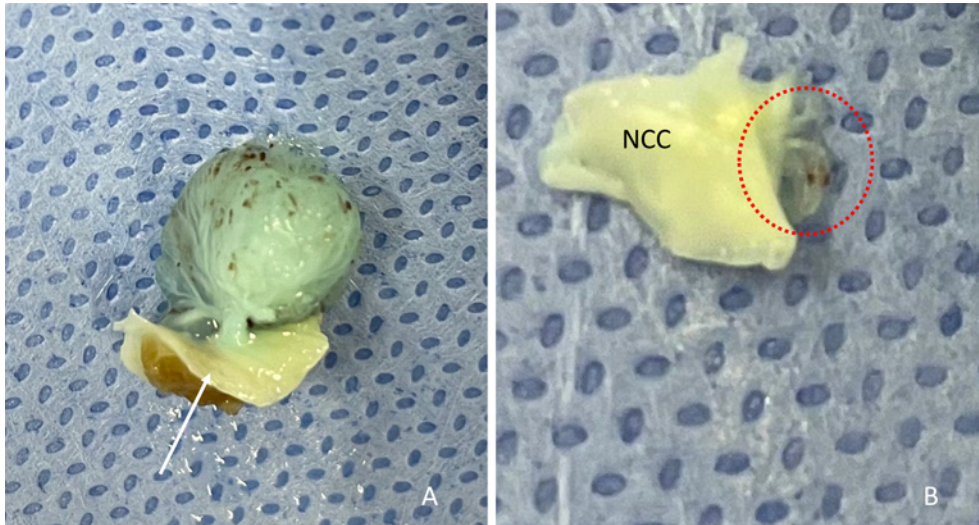
**Figure 1.** **A:** Diffusion-weighted magnetic resonance imaging demonstrated hyperintense signal in the right corona radiata. **B:** Diffusion-weighted magnetic resonance imaging demonstrated hyperintense signal in the right parietal lobe. **C:** Magnetic resonance angiography revealed the right middle cerebral artery was occluded at the point indicated by the red arrow.



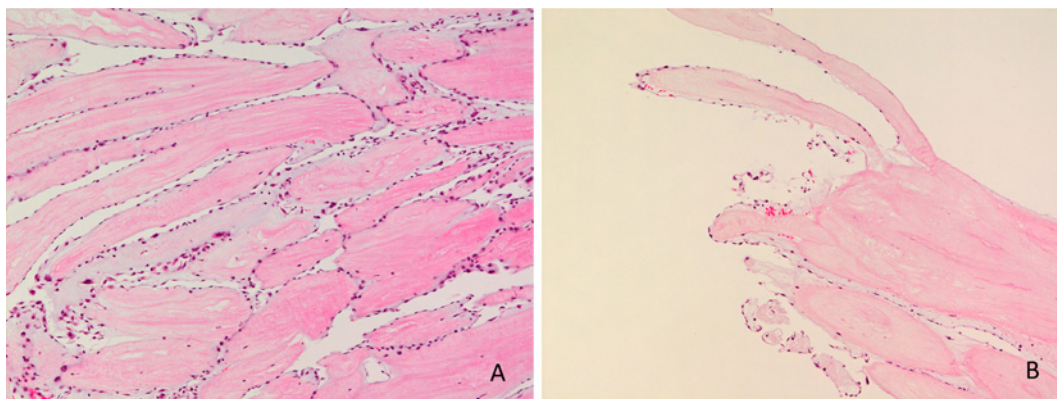
**Figure 2.** Transesophageal echocardiography. **A:** A mobile round mass (white arrow) with a stalk attached to the septum in the left ventricular outflow tract. **B:** A mobile strand (encircled) attached to the noncoronary cusp. LV indicates left ventricle; LA, left atrium; and Ao, aorta.

phy was normal. Transthoracic echocardiography (TTE) demonstrated an ejection fraction of 79%, no valve disease, and a hyperechoic spherical mass moving with ventricular contraction in the LVOT. Transesophageal echocardiography (TEE) revealed a pedunculated mass with a narrow stalk attached to the left ventricular septum (Figure 2A). The mobile mass was  $10.2 \times 7.7$  mm in size without significant elevation of the LVOT Doppler velocity. Enhanced computed tomography (CT) demonstrated no tumor-like mass or embolism in any other organ. Cardiac MRI revealed that the mass was isointense in T1-weighted images and hyperintense in T2-weighted images. A perfusion delay was observed only in the mass but not in the left ventricular wall. Following gadolinium injection, enhanced imaging confirmed that late inhomogeneous gadolinium enhancement was present in the mass. Coronary CT showed no significant stenosis in the coronary arteries. This ischemic stroke was caused by embolism from the mass in the LVOT. After 14 days of conservative treatment, his neurological symptoms gradually improved, and 2 months later, surgery was scheduled. The surgery was performed under cardiac arrest with cardiopulmonary support. The ascending aorta was resected

transversely, and the tumor was transaortically approached, which appeared to be a sea anemone-like feature with a stalk. Further inspection revealed a small filiform lesion on the outer leaflet of the noncoronary cusp (NCC). The lesion was similar to that of the mass in the LVOT and stuck to the NCC. First, the tumor in the LVOT was resected with the myocardium. The gross specimen was  $11 \times 7 \times 6$  mm in size (Figure 3A). After the removal of the filiform lesion, a small fenestration was left in the NCC. The hole was too large for valvuloplasty, leading to replacement of the aortic valve. The latter lesion was 3 mm in size (Figure 3B). Pathologic analysis revealed that the mass in the LVOT consisted of a fibrous connective tissue core and frond-like branches covered by multiple layers of endothelium, which is consistent with the characteristics of PFE (Figure 4A). In contrast, the latter lesion consisted of fibrous connective tissue covered by a single thin endothelium and was diagnosed as LE (Figure 4B). His postoperative course was uneventful, and he was discharged without complications. We retrospectively checked the TEE prior to surgery and found a thin mobile mass on the aortic valve, which was considered LE (Figure 2B). However, the latter mass of the NCC was



**Figure 3.** Gross appearance of the masses in the left ventricular outflow tract (LVOT) and on the noncoronary cusp (NCC). **A:** The mass of LVOT resembled a sea anemone with a short stalk attached to the myocardium (white arrow). **B:** The filiform lesion (encircled) located in the outer surface of the NCC.



**Figure 4.** Hematoxylin-eosin stain, magnification  $\times 100$ . **A:** A pathological study of the mass on the left ventricular outflow tract demonstrated thick frond-like branches, called a fibro-vascular core, covered by multiple layers of endothelium. **B:** A pathological study of the filiform lesion on the noncoronary cusp demonstrated thin vascular cores covered by single flat endothelium.

not found on MRI or CT retrospectively.

### Discussion

We report the rare case of coexisting PFE in the LVOT located at an unusual site and incidentally found LE during the surgical procedure. We retrieved the preceding article in PubMed, and a case of stroke associated with the left heart associated with both papillary fibroelastoma and Lambl's excrescence was not found. Careful inspection via TEE should be performed for patients who are suspected to suffer embolic stroke. PFEs are detected incidentally or while searching for causes of embolism. TTE should be the first option for detecting PFE. Early detection of PFE using TEE was described by Gowda, *et al.*<sup>2)</sup> Sun, *et al.* studied echocardiographic characteristics, which included a round, oval or irregular appearance with

well-demarcated borders and a homogenous texture. The size ranged from 2 to 28 mm in the largest dimension. Of 110 tumors, 48 (43.6%) had stalks and were easily mobile. The sensitivity and specificity of TTE were 88.9% and 87.8%, respectively, for the detection of PFE greater than 0.2 cm in size.<sup>4)</sup> In our case, the tumor was a mobile, round-shaped mass with a stalk and was approximately 10 mm in size. The features were typical, and the size was large enough, leading to easy identification of PFE by TTE. Tamin, *et al.* showed that the cerebrovascular accident (CVA) risk was 6% at 1 year and 13% at 5 years, which was greater than that of patients who underwent surgery. These findings suggested that PFE should be considered for resection for the prevention of CVA in patients who are good surgical candidates and who have left-sided PFE, regardless of size, mobility or location.<sup>3)</sup> In addition, Sakaguchi, *et al.* resected PFE in the LVOT by opening

the ascending aorta and inspecting through the aortic annulus without any difficulties because the PFE was close to the aortic annulus. PFE is fragile and can break into pieces during resection, possibly leading to embolization during surgery, requiring careful resection.<sup>6)</sup> Patients who are not surgical candidates could be treated with oral anti-coagulation agents.<sup>2)</sup> In our case, the patient had symptoms and was a good surgical candidate for surgery. TEE showed normal valve function, and the tumor was close to the aortic annulus and could be resected transaortically. During surgical inspection, the tumor on the outer surface of the NCC was treated like the tumor in the LVOT. We should emphasize that the fragile tumors on the NCC are at risk of embolism. LE is a filiform valvular frond < 2 mm in thickness and > 3 mm in length and is located mostly on the valve closure line of the mitral valve and the ventricular side of the aortic valve. LE can also cause thromboembolic events. TEE is recommended to evaluate the cause of the stroke, and LE is found incidentally. LE appears as an undulating, hypermobile, strand-like structure at the site of valve closure.<sup>3)</sup> However, the sensitivity and specificity of detection of mobile echogenic structures by TEE are 68% and 85%, respectively, mainly because standardized definitions for identifying mobile echoes are insufficient.<sup>7)</sup> The differences between PFE and LE are as follows: PFE is larger, often solitary, arising from the midportion of valve leaflets, and possibly arising from less mechanically affected areas, including the endocardium. On the other hand, LE usually occurs in multiples forms and arises from the coaptation of valve leaflets.<sup>3)</sup> In our case, the tumor on the NCC was found during surgery and was not detected by TEE before surgery. In addition to the low values of detection by TEE, the PFE in the LVOT led us to overlook the LE because the PFE was obviously considered to be the cause of the embolism. To the best of our knowledge, there are no reports of MRI or CT findings that play a role in identifying LE. In this patient, LE was not found by MRI or CT. It is possible that the LE was undersized because parts of the LE were embolized. We cannot clarify which of these conditions caused stroke, PFE or LE. According to the diameter of the right middle cerebral artery, which was approximately 3 mm, the stroke was presumed to be caused by a larger PFE. Asymptomatic patients with LE should be followed by TEE instead of surgical resection.<sup>3)</sup> If LE is found before surgery, we could consider whether LE should be resected in terms of the probability of stroke. When we incidentally found LE during surgery, we did not have enough

time to consider this phenomenon. We thought that LE should also be resected because of the possibility of stroke. If LE is not detected before surgery, LE is likely to be overlooked during surgery. However, surgical inspection for LE is not needed because LE occurs in 70-80% of adult heart valves<sup>8)</sup> and asymptomatic LE does not need to be resected.<sup>3)</sup> Therefore, careful inspection via TEE before surgery should be performed to determine treatment.

## Conclusion

We have reported a rare case of PFE in the LVOT, which is an unusual location, and LE on the NCC was incidentally found during surgery. A careful inspection should be performed because PFE can cause us to overlook other significant causes.

## Disclosure

**Conflicts of interest:** The authors declare there are no relationships relevant to this paper and that there are no conflicts of interest to disclose.

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