Lambl’s Excrescences Associated with Cardioembolic Stroke

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Patient: Male, 73-year-old
Final Diagnosis: Lambl’s excrescence • stroke
Symptoms: Blurring of vision • weakness
Medication: —
Clinical Procedure: —
Specialty: Cardiology • Neurology

Objective: Rare disease
Background: Lambl’s excrescences (LE) are threadlike fronds that occur along valve closure lines where minor endothelial damage occurs, resulting in thrombus formation. It is often asymptomatic but can result in cerebral embolism and coronary artery obstruction. The criterion standard for diagnosis is transesophageal echocardiography.

Case Report: We report an interesting case of a 73-year-old right-handed man presenting with a visual disturbance. An MRI head demonstrated an acute-to-subacute infarct in the right posterior cerebral artery territory involving the posterior right temporal, right occipital lobes, and right thalamus, which was in keeping with embolic stroke. Investigation with transthoracic and transesophageal echocardiography showed echo density on the ventricle surface of the left coronary cusp, which was concerning for Lambl’s excrescences, with no significant arrhythmia on his implantable loop recorder at 3-month follow-up. Therefore, we believe that his stroke was due to embolization from Lambl’s excrescences. Since this was his first episode of stroke, monotherapy with aspirin was continued.

Conclusions: Although rare, Lambl’s excrescences should be considered in the differential diagnosis of embolic stroke. However, there is no established guideline for its management. Patients with the first stroke episode can be treated conservatively with antiplatelet therapy. In patients with recurrent ischemic events, anticoagulation should be offered, along with discussion about surgical excision.

Keywords: Echocardiography, Three-Dimensional • Embolic Stroke • Infarction, Posterior Cerebral Artery

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Background

Cerebrovascular accident (CVA) is a leading cause of morbidity and mortality globally. However, among the 87% of ischemic stroke cases, cardiac embolism accounts for only 14-30% of all cases [1]. On the other hand, cryptogenic stroke represents 24-28% of all ischemic strokes, according to a meta-analysis reported by Ornello et al [2]. Lambl’s excrescences (LE) are a rare cause of embolic stroke, first described in 1856 by the Bohemian physician Vilém Dušan Lambl [3]. These are branched filiform structures with undulating movements on aortic and mitral valves. These are more common on the atrial side of the mitral valve (68-76%) followed by the ventricular side of the aortic valve (38-50%) [4]. Their pathogenesis remains uncertain and is believed to result from valvular wear and tear and have been considered to be part of normal aging. However, the clinical significance and effective treatment of LE remain controversial [3]. Here, we present an interesting case of a 73-year-old man presenting with visual deficits, found to have ischemic stroke thought to be secondary to LE.

Case Report

A 73-year-old right-handed man with a history of poorly controlled type 2 diabetes mellitus (HbA1C 10.4) on home insulin therapy presented to the Emergency Department (ED) after a fall associated with lightheadedness and nausea for 1 day. He also mentioned having visual disturbances in left upper visual fields for the same duration. Vitals were stable, and examination revealed a dense left homonymous hemianopsia and abnormal (ataxic) left finger-to-nose test. “Stroke alert” was called. Non-contrast computed tomography scan (CT) of the brain revealed acute right posterior cerebral artery distribution infarct (Figure 1).

He was not a candidate for intravenous tissue plasminogen activator as his last known well was greater than 4.5 h. CT angiogram of the brain again showed ischemia in the right posterior cerebral artery distribution, with the suggestion of a right occipital and temporal lobe core infarct. CT angiogram of the neck did not show any hemodynamically significant vessel stenosis. Ultrasound Doppler of bilateral carotids was done, which showed plaque formation within both carotid bulbs, with 40% diameter stenosis. He was started on antiplatelet therapy with aspirin (ASA) and high-intensity statin. MRI angiography head (Figure 2) showed restricted diffusion in the right posterior cerebellar artery (PCA) territory infarction involving the posterior right temporal and right occipital lobes.

Figure 1. CT head without contrast with black arrows showing decreased density representing acute right posterior cerebral artery distribution infarct.

Figure 2. Axial diffusion-weighted MRI brain without contrast, with black arrows demonstrating acute-to-subacute right posterior cerebellar artery (PCA) territory infarction involving the posterior right temporal and right occipital lobes.
Lambl's excrescences (LE) are most commonly found on the atrial surface of the mitral valve (68-76%) and the ventricular surface of the aortic valve (38-50%) and have been rarely described on native right-sided valves and prosthetic valves [5].

**Discussion**

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The patient had echo density on the left coronary cusp in our case. These are primarily asymptomatic and may present as an incidental finding on echocardiography. However, LE can break off, as in our case, and lead to thromboembolic events. Ischemic events have been more reported with involvement of the aortic valve, and no correlation has been found between the size of strands and the incidence of stroke. Valvar strands can be detected on high-resolution transthoracic echocardiography (TTE), which appears at the leaflet coaptation as hypermobile, undulating, strand-like structures less than 2 mm thick more than 3 mm long along the valve closure [5]. Differential diagnosis includes imaging artifact, vegetations, thrombi, redundant leaflet, flap due to aortic dissection, papillary fibroelastoma, myxoma, other cardiac neoplasms, and metastases. Fibroelastomas are the most difficult to differentiate from LE. Notable differences from LE are multiple lesions and locations along the lining of the valve closure.

Fibroelastoma is solitary and larger, usually arising from the midportion of the valve leaflet, which at times can appear as a pedunculated lesion. In our case, the echocardiogram showed a low-density, thin, hypermobile, filiform lesion on the ventricular surface of the aorta along with the valve closure, consistent with LE. As in this case, high-resolution TTE can diagnose LE, but the criterion standard for diagnosis is transesophageal echocardiography (TEE), which can best define the lesion [4]. There is no standard evidence-based guideline in LE management, given the condition’s rarity. Management is based mainly on case reports and includes single/dual antiplatelet agent vs anticoagulation vs surgery [1]. An asymptomatic patient can be monitored closely and followed with transesophageal echocardiography (TEE). Chu et al suggest that LE should be considered in the differential diagnosis of cryptogenic stroke and recommend performing TEE [6]. Patients with the first episode of cerebrovascular accident can be treated with antiplatelets, with some authors advocating aspirin alone [7,8] and others advocating dual antiplatelet agents [6,9]. In our case, we used aspirin monotherapy, as this was his first episode of stroke. The recommendation of whether to use anticoagulation versus antiplatelet in patients with a single embolic event is unclear [10,11]. If there is a recurrence of the ischemic events while on antiplatelet therapy, the addition of anticoagulation therapy with warfarin should be considered before proceeding with surgical intervention [6]. It has a recurrence rate of around 30% [1] CVA after the second episode of a thromboembolic event. Hence, it is reasonable to offer surgical intervention with shared decision-making with the patient [12].

**Conclusions**

We recommend LE be considered in the differential diagnosis of embolic stroke. Patients with a first stroke episode can...
be treated conservatively with antiplatelet therapy. Shared decision-making about anticoagulation and surgical excision should be done in patients with recurrent ischemic events. In patients with poor or doubtful compliance with medication, surgery can be considered as the primary treatment.

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