

Lambl's Excrescences – Taking the Crypto out of Cryptogenic Stroke: A Case Report and Literature Review

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Abstract Lambl's excrescences (LE) are thin and long filiform projections arising from the line of closure of cardiac valves. Their etiology is not well understood, but they're thought to result from a wear-and-tear process. They are typically asymptomatic and best detected by transesophageal echocardiogram. Although controversial, some studies suggest an association between LE, ischemic stroke, and headaches. We describe a 56-year-old man who presented with a 3-day history of bilateral blurry vision within the left visual field. One day prior to his visual symptoms, he developed a severe headache that persisted until his presentation to our hospital. On physical examination, visual acuity and fields were intact, but he reported blurriness in the left visual field of both eyes. Brain MRI showed a wedge-shaped area of restricted diffusion along the medial aspect of the right occipital lobe, indicating an ischemic stroke of the visual cortex. Due to the embolic appearance of the stroke, transesophageal echocardiogram was performed and showed a LE on the aortic valve. No other embolic source was identified. In the absence of another explanation for his stroke, LE was considered the most likely source of his embolic stroke. The patient was treated with antiplatelet therapy. In conclusion, this case report highlights LE as a possible cardioembolic source of stroke and discusses its association with headache. We also provide a review of the current literature on LE and the lack of clear guidelines on its management.

Keywords: Cerebrovascular disease, Stroke, Embolism, Cardiac, Cardioembolic stroke, Lambl's Excrescences, Headache

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1. Introduction

Lambl's excrescences (LE) are defined as fine, mobile, filamentous extensions of the cardiac valves. The etiology of LE is not well understood, but it's thought to result from a wear-and-tear process. [1] LE are typically asymptomatic and discovered incidentally. Stroke can be classified as either ischemic or hemorrhagic with ischemia accounting for most of the events. [2] Cardiogenic embolism accounts for almost 20% of ischemic strokes. Etiologies of cardioembolic stroke include valvular disease, atrial fibrillation, left ventricular thrombi, left atrial thrombi, and cardiac tumors (e.g., atrial myxoma). Although extremely rare, LE has been described in several case reports as the culprit for thromboembolic events. [3]

2. Case Presentation

We describe a 56-year-old man who presented with bilateral blurry vision for 3 days. He described "rainbows and stars" along with some flashes moving within his left visual field that lasted a few seconds and disappeared with eye movement. The visual symptoms recurred if he focused his vision on something. One day prior to the onset of his visual symptoms, he started to complain of a new-onset headache originating from the right frontal region and radiating to the right temporal region. The headache was severe, continuous, and throbbing in nature. His medical history was notable for hyperlipidemia and a 45-pack-year smoking history. His neurological examination was non-focal. Visual acuity and fields were intact, but he reported blurriness in the left visual field bilaterally. Brain magnetic resonance imaging showed a wedge-shaped area of restricted diffusion along the medial aspect of the right occipital lobe, indicating an ischemic stroke of the visual cortex, as shown in Figure 1.



Figure 1. A brain MRI showing a wedge-shaped area of restricted diffusion along the medial aspect of the right occipital lobe

Computed tomography angiogram of the head and neck were negative for any bleeding or significant stenosis of the neck or intracranial arteries. Laboratory testing revealed a total cholesterol level of 233 mg/dL and an LDL cholesterol level of 173 mg/dL. His comprehensive metabolic panel, hemoglobin A1C, coagulation profile, complete blood count, c-reactive protein, and erythrocyte sedimentation rate were all normal. An initial 12-lead electrocardiogram (ECG) and subsequent 3-day telemetry monitoring showed a sinus rhythm. Transthoracic echocardiogram (TTE) revealed normal left and right ventricular function and size, and no valvular dysfunction. Due to the embolic appearance of his stroke, a transesophageal echocardiogram (TEE) was performed and revealed a small fibrinous strand (i.e., Lambl's exrescence) on the left ventricular outflow tract of the aortic valve, as shown in Figure 2. TEE also showed no patent foramen ovale, no left atrial appendage thrombus, and a mild localized sessile atheroma measuring 0.1 cm in the mid-descending thoracic aorta. The left atrial cavity was normal in size, and the left atrial appendage was not multilobed.

A supplemental video (video 1.) is provided.



Figure 2. A TEE still picture showing a thin linear structure arising from the aortic valve

The presence of Lambl's exrescence in this patient suggested a possible embolic source of his stroke,

particularly in the absence of another explanation.

The patient was educated about the paucity of data on the optimal therapy for LE. He was offered antiplatelet and anticoagulation therapy, and he opted for the former. He was started on atorvastatin and aspirin, and smoking cessation was strongly recommended. After discharge, a 30-day event monitor was completed and did not show any arrhythmias. One month after discharge, he was seen in the cerebrovascular clinic and no additional events had occurred at that time.

3. Discussion and Conclusion

LE typically show undulating independent motion and are more common on the mitral valve than the aortic valve. Although often confused with papillary fibroelastomas, LE appears as a thread-like strand arising from the line of closure of a valve on TEE. In contrast, a papillary fibroelastoma typically look like a pedunculated "frond-like" tumor arising from the mid-portion of valve leaflets. [4]

In a study that enrolled 619 stroke patients, valvular strands were present in 244. A total of 265 patients were classified as having cryptogenic stroke. [5] Another study suggested a particularly increased risk of ischemic stroke among younger patients with LE. LE of both the mitral and aortic valves were associated with cerebral ischemia, with a slightly higher risk in patients with aortic LE. [6] On the other hand, some studies suggest no increased risk of cardioembolic strokes in patients with LE. [7]

Headache, as was seen in our patient, is sometimes seen with stroke (including ischemic stroke), and the etiology remains unclear. An association between LE and headache has not been established, but Liu et al. described two patients with LE who presented with a migraine-like headache and ischemic stroke. [8] The authors hypothesized that the headaches might have been triggered by cerebral ischemia due to micro-emboli originating from LE. Davogustto et al. described another patient with LE who had migraine-like headaches. [9] While definitive conclusions about our patient's headache cannot be drawn, the temporal association between the headache onset and subsequent development of his ischemic stroke could be explained by micro-emboli originating from his LE.

Although there are no clear guidelines for the management of LE, some experts recommend close follow-up with serial echocardiograms for asymptomatic patients with LE. [10] For patients with LE who have experienced a stroke with no alternative source of emboli, treatment with anticoagulation, or aspirin and clopidogrel has been suggested. [10] For patients with LE who experience recurrent strokes while on anticoagulation, a surgical debridement of the LE may be considered [10].

In conclusion, LE should be considered as a possible embolic source in otherwise cryptogenic strokes. TEE is superior to TTE in identifying LE and should be included in the work-up of a cryptogenic stroke. While the etiology of headache in ischemic stroke remains unclear, cerebral ischemia due to micro-emboli might play a role. No definitive guidelines direct management of LE currently, so more studies are needed to clarify the best approach.

A supplemental table (Table 1) summarizes previously reported cases of LE.

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Year of the case report	Age	Gender	Presentation	LE's Location	Management	
1981 [11]	54	Female	Incidental finding	Papillary muscle and aortic valve	Surgical excision	
1982 [12]	70	Male	Embolus to the left popliteal artery	Unknown	Echocardiographic surveillance	
1995 [13]	31	Female	Ischemic stroke	Mitral valve	Surgical excision Acenocoumarol	
1996 [14]	64	Female	Angina	Aortic valve	Surgical excision Aspirin 250 mg	
1999 [15]	72	Male	Ischemic stroke	Pulmonary valve	Unknown	
1998 [16]	44	Female	Weakness and fatigue	Aortic valve	Surgical excision	
2003 [1]	66	Female	Ischemic stroke	Aortic valve	Anticoagulation Surgical excision	
2006 [17]	47	Female	Ischemic stroke	Aortic valve	Aspirin 100 mg	
2006 [18]	42	Male	Transient ischemic attack	Aortic valve	Aspirin 200 mg	
2007 [19]	61	Female	Ischemic stroke	Aortic valve	Surgical excision	
2007 [20]	80	Female	Incidental finding	Aortic valve	Unknown	
2008 [21]	69	Female	Incidental finding	Aortic valve	Surgical excision	
2010 [22]	59	Male	Ischemic stroke	Aortic valve	Warfarin	
2012 [23]	50	Female	Myocardial infarction	Aortic valve	None	
2012 [8]	53	Male	Ischemic stroke	Aortic valve	Aspirin 100 mg	
2012 [8]	30	Male	Ischemic stroke	Aortic valve	Surgical excision	
2012 [24]	N/A	Female	Incidental finding	Aortic valve	Surgical excision	
2013 [25]	66	Female	Ischemic stroke	Aortic valve	Warfarin	
2013 [26]	33	Male	Ischemic stroke	Mitral valve and chordae tendineae	Surgical excision	
2014 [27]	59	Male	Ischemic stroke	Aortic valve	Aspirin	
2015 [9]	68	Female	Ischemic stroke	Aortic valve	Aspirin	
2015 [3]	51	Female	Two ischemic strokes and one trainset ischemic attack	Aortic valve	Aspirin 81 mg Clopidogrel 75 mg	
2015 [28]	64	Female	Viral myocarditis	Aortic valve	Anti-platelet therapy	
2015 [29]	48	Male	Ischemic stroke	Aortic valve	Unknown	
2016 [30	74	Female	Ischemic stroke	Aortic valve	Aspirin	
2016 [31]	42	Male	Transient ischemic attack	Aortic valve	Anticoagulation	
2016 [32]	59	Male	Myocardial infarction	Aortic valve	Surgical excision	
2018 (our case report)	56	Male	Ischemic stroke	Aortic valve	Aspirin 81 mg	

Conflict of Interest

The authors declare that they have no conflict of interest.

List of Abbreviations

LE: Lambl's exrescence;

- TTE: transthoracic echocardiogram;
- TEE: transesophageal echocardiogram;

ECG: electrocardiogram

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