

Child Neurology: Case Report of Lambl Excrescences in a Pediatric Patient With Multifocal Strokes

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Abstract

Lambl excrescences are fibrinous connective tissue strands found predominantly on left-sided cardiac valves. These valvular strands are typically benign, but have been implicated as a potential etiology of embolic strokes in adult patients. The significance of Lambl excrescences in pediatric stroke cases is unclear and not previously reported in the literature. In this study, we describe a 10-year-old boy who presented with acute-onset right-sided hemiplegia, found to have multifocal embolic strokes of various ages. Extensive stroke workup was unrevealing, aside from the presence of small, filamentous, strand-like densities associated with the mitral and aortic valves noted on a transesophageal echocardiogram consistent with Lambl excrescences. In this case report, we review Lambl excrescences and their significance in acute stroke and management options for the prevention of future ischemia in these patients.

Clinical Case

An obese 10-year-old boy presented with acute-onset right-sided hemiplegia. Several days earlier, he experienced mild abdominal discomfort without vomiting, diarrhea, fever, and other illness symptoms. On the day of presentation, he was fatigued and slept most of the day. By afternoon, his parents noticed that he had a right facial droop, slurred speech, difficulty walking, and confusion. In the emergency department, he was found to have right-sided hemiplegia. NIH stroke score was 8 on presentation. A CT angiogram of the head and neck showed an acute left middle cerebral artery (MCA) stroke with filling defects in the left post bifurcation M1 segment and the left M2 segment vessels. In addition, the imaging was consistent with chronic ischemia in multiple vascular territories, including the right posterior MCA, left MCA, and left posterior cerebral artery (PCA), with right parietal and left occipital encephalomalacia suggestive of remote injury. MRI confirmed acute left MCA territory ischemia with involvement of the left caudate and putamen and the presence of previous infarcts visualized on CT (Figure 1). Of note, the family did not report past episodes suggestive of symptoms related to previous infarcts. Given an overall clinical picture, he was not considered to be a candidate for recanalization therapies. He was given 325 mg of aspirin and placed on 81 mg of aspirin daily.

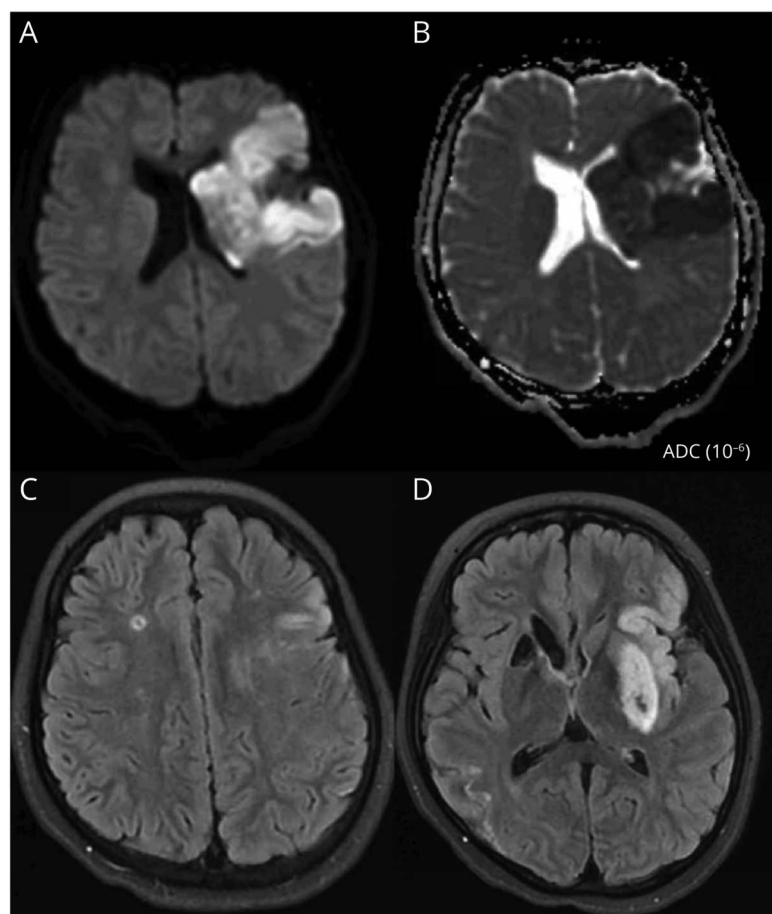
He underwent an extensive stroke workup. Complete blood count, comprehensive metabolic panel, hemoglobin A1c, cholesterol panels, COVID-19 PCR and antibody, erythrocyte sedimentation rate, C-reactive protein, antinuclear antibody, antineutrophil cytoplasmic antibodies, antiphospholipid panel, and inherited thromboses panels were all normal. Vessel wall imaging demonstrated eccentric enhancement at the site of emboli previously visualized in the left M1 and M2 segments, consistent with known thrombus. No other vessel wall abnormalities or features concerning for vasculitis were seen. An ECG finding was normal. He had no history of arrhythmias or palpitations. A transthoracic echocardiogram (TTE) with agitated saline bubble

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(A) Diffusion-weighted imaging with diffusion restriction in the left middle cerebral artery territory. (B) Apparent diffusion coefficient correlating to area of diffusion restriction. (C and D) T2 fluid-attenuated inversion recovery demonstrating areas of chronic infarct.

study showed normal cardiac structure and function, intact septae, and no evidence of intracardiac masses or vegetations. He did not have any recurrence of abdominal pain or other symptoms that would suggest systemic disease. There was no family history of strokes or hematologic disorders.

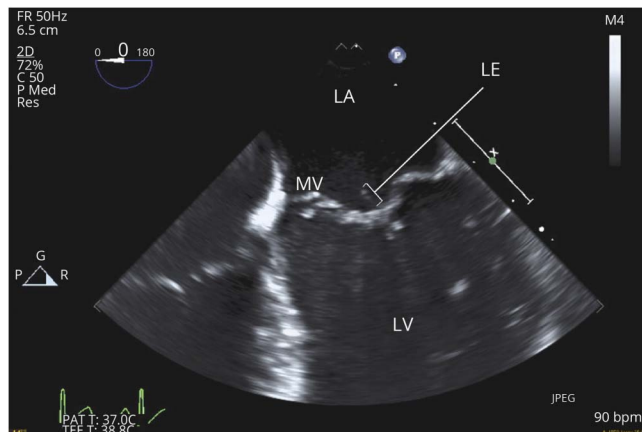
A cardioembolic etiology remained a concern, given his strokes of varying ages in multiple vascular territories. A transesophageal echocardiogram (TEE) was performed because of suboptimal transthoracic imaging in an obese child (body mass index 31.8 kg/m², 99%). TEE revealed several small, linear, strand-like mobile densities on the atrial aspect of the mitral valve (Figure 2) and the ventricular aspect of the aortic valve consistent with Lambl excrescences (LEs). No additional masses, vegetations, or evidence of thrombi were found within the atria, atrial appendages, or ventricles. There was no suspicion for endocarditis. Given these findings, he was placed on dual antiplatelet therapy with aspirin and clopidogrel indefinitely. During discharge, symptoms had improved, and he had residual mild right-sided weakness and coordination difficulties. Repeat MRI 2 months later demonstrated evolution of acute infarct without any new strokes, and he has had no further episodes concerning for ischemia.

Discussion

Role of Echocardiography in Arterial Ischemic Stroke Evaluation

The incidence of non-neonatal pediatric stroke is estimated to be at least 1–2/100,000 children annually with approximately one-third of these from cardioembolic etiologies, often from known cardiac disease.^{1,2} TTE with bubble contrast study and ECG are generally obtained as part of a pediatric ischemic stroke workup. TTE is usually adequate for the identification of most structural abnormalities in children, including many intracardiac vegetations. TEE is less commonly obtained because of its more invasive nature and the need for sedation; however, it has been suggested as an additional diagnostic tool if TTE is unrevealing and the diagnosis otherwise remains elusive.³ In particular, TEE should be considered when transthoracic imaging is suboptimal (e.g., chest wall deformities, previous surgery, obesity, etc). TEE may yield additional findings not detected by TTE in up to half of patients with embolic strokes of undetermined source (ESUS) in the adult population.⁴ In our patient, TEE was indicated due to the presence of multiple strokes of various ages in multiple distributions without another identifiable etiology and to ensure adequate imaging

Figure 2 TEE Still-Frame Image of the Patient's Mitral Valve Showing an Approx. 5 mm Thin, Filamentous Strand on the Atrial Aspect of the Mitral Valve Consistent With LE



LA = left atrium; LE = Lambl excrescence; LV = left ventricle; MV = mitral valve; TEE = transesophageal echocardiogram.

of the valves in an obese child with less-than-optimal transthoracic acoustic windows.⁵

Significance of LEs

LEs are mobile, filiform, fibrinous, connective tissue strands that occur at and around the coaptation site of cardiac valves: a wear-and-tear lesion. They are most frequently found on the atrial aspect of the mitral valve and ventricular aspect of the aortic valve with rare right-sided valvular involvement.⁶ Histologically, they are composed of a connective tissue center with collagen and elastic fibrils enclosed by a layer of endothelium.⁷ These valvular strands are typically benign findings and rarely require management or intervention. There is no known association between these lesions and connective tissue disorders, genetic variations, or specific valvular injury. They are quite rare in children in general, with 1 study showing a prevalence of approximately 1.7% and mean age of 11 ± 5.9 years (range 5 months–17 years).⁸ The reported prevalence in adults is somewhat variable but seems to increase with age with reported rates of LEs varying from 5.5% across all adults to as high as 56% in adults 80–90 years of age.^{6,9}

The differential diagnosis for LEs includes infective endocarditis, thrombus(i), atrial myxomas, papillary fibroelastoma, and imaging artifact. Papillary fibroelastomas are benign neoplastic growths of the cardiac valves and are particularly difficult to distinguish from LEs because they are histologically similar but do have some key characteristics that allow them to be differentiated on echocardiography: they are typically solitary in occurrence, arise from the middle of the valve, and are more pedunculated in nature. Infective endocarditis typically presents with symptoms of intermittent fevers, fatigue, and secondary immune phenomenon in addition to embolic phenomenon. Endocarditis is rare in the pediatric heart in the absence of previous cardiac surgery,

rheumatic heart disease, or indwelling central lines.¹⁰ Atrial myxomas typically are more mass-like and can occur anywhere within the atria, but most commonly the left atrium, particularly at the mitral annulus or fossa ovalis.¹¹ Thrombus likewise can occur anywhere and is not typically limited to the cardiac valves.

Literature regarding the association of LEs and stroke is somewhat conflicting and limited to adult cases. Studies of retrospective TTE and TEE data in adults have shown a significant association between LEs and embolic disease,^{9,12} and adult case reports have suggested an association between LEs and cardioembolic stroke.¹³ Conversely, a study of adults younger than 60 years comparing prospective TEE data without stroke with retrospective TEE data of those with ESUS showed no significant association between LEs and stroke.⁶ In our review of literature, there is essentially no data or reports of children with cryptogenic stroke who have been found to have LEs.

Stroke Recurrence Prevention in Patients With LEs

There are currently no accepted standard practices or guidelines for the management of stroke prevention when LEs have been detected. Recommendations in the adult literature vary from no treatment in asymptomatic patients to consideration of surgical excision in recurrent strokes.¹⁴ One recent review of the adult literature proposed an algorithm that recommends stroke prevention in patients with known stroke or TIA in the setting of LEs¹⁵ with initial treatment consisting of dual antiplatelet therapy (aspirin and clopidogrel) vs anticoagulation, while reserving surgical debridement for failure of either or both treatments with recurrent strokes. Timing and option for surgical debridement should be thoroughly discussed with patients along with the associated risks and benefits.

Given the paucity of pediatric literature, the optimal management of stroke prevention in children with embolic stroke and LEs is unclear. Owing to the multifocal, subclinical strokes of various ages in our patient with no other identifiable source, we opted to start dual antiplatelet therapy using aspirin and clopidogrel for stroke prevention indefinitely. Our patient has not had recurrence of stroke since the initiation of antiplatelet therapy. In pediatric patients with stroke and LEs, other etiologies should be evaluated thoroughly, with consideration of LEs as the etiology of stroke after exclusion of other causes. The clinical course of individual cases should guide treatment decisions, and the risks/benefits of stroke prevention options should be discussed thoroughly with families. Repeat cardiac imaging may also help guide management, especially in situations of repeat stroke.

Conclusion

LEs are filamentous, fibrinous strands on left-sided cardiac valves because of wear and tear and are typically incidental findings. Literature in adults regarding their association with embolic stroke is conflicting. To our knowledge, this is the first reported case of embolic stroke potentially related to LEs

in a child. Although it is possible our patient had additional risk factors contributing to stroke, given most LEs are asymptomatic, none were identified after extensive evaluation, and he has not had additional strokes since the initiation of dual antiplatelet therapy. LEs should be considered in the differential for pediatric patients with embolic strokes, and in some cases, TEE should be considered in the evaluation. Dual antiplatelet therapy or oral anticoagulation is a reasonable starting point for the prevention of recurrent stroke before considering surgical debridement.

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Disclosure

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Appendix (continued)

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