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Lambl's filaments as a cause for cardiogenic embolism of cerebral vessels and coronary arteries

Presented by Academician of the NAS of Ukraine V.P. Shyrobokov

The article presents a specific clinical case and literature data on a rare finding in the heart cavity – Lambl's filaments. Histologically, these are cells of fibroelastic tissue covered with stroma and epithelial layer. They do not have a vascular sheath, so they are devoid of granulation tissue. Lambl's growths occur on both their own and prosthetic heart valves. Very rarely, threads can also be seen in areas other than heart valves such as the papillae, atria, and chords. These lesions can be seen on the echocardiogram accidentally or in connection with serious complications such as embolic stroke or acute coronary syndrome after their separation and distribution to distant organs. Lambl's growths occur in adults and children, but the prevalence is higher in the former and increases with age. On the echocardiogram, the incidence reaches a peak between the ages of 61 and 70, and then decreases due to the aging of heart valves, which are increasingly calcified. The disease is more common in men than in women.

Keywords: *Lambl's filaments, stroke, angina.*

Lambl's filaments are a histological term that describes rare heart growths that develop on heart valves and are not considered to be thin, hypermobile, filamentous fibrils. These lesions may be seen on echocardiography accidentally or due to serious complications such as embolic stroke or acute coronary syndrome afterwards [1]. Lambl's filaments occur as small growths on the endocardial surfaces and have the potential to embolize distant organs. Histologically, these are cells composed of a core made of fibroelastic tissue covered with a stroma, which is densely hyalinized. This is covered, in turn, by a single layer of endothelium. They do not have a vascular membrane, so they are devoid of granulation tissue. Lambl's growths are found on both own and prosthetic heart valves. Very rarely, filaments can also be seen in areas other than heart valves, such as papillary muscles, atrial septa, and chords [2, 3].

Lambl's threads appear as individual strands and in clusters with a diameter of about 1 mm and a length of 10 mm or more. They can also lengthen due to their location on the valve leaflets, which are subjected to heavy loads, especially if they are located on the aortic valve, which

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is subjected to a relatively greater pressure than others. Sometimes, multiple adjacent filaments are adjacent to one another, by forming “giant Lambl's growths” that can grow up to 2 cm. Despite some cases where giant Lambl's growths are associated with ischemic stroke, there is no clear evidence in the medical literature of a correlation between strand size and the risk of an embolic event [4].

Epidemiology. In the general population sent for transesophageal echocardiography, 5.5 % of people have valve threads, and the number increases sharply to 40 % in patients who have suffered a cryptogenic stroke. Lambl's growths are found in adults and children, but the prevalence is higher in the former and increases with age. By echocardiography, the prevalence peaks between the ages of 61 and 70, and then decreases due to the aging of the heart valves, which become more and more calcified. The disease is more common in men than in women. Even though Lambl's growths may be present on all native and prosthetic valves, the prevalence is higher on the left heart valves (more common on the mitral than on the aortic one) [5, 6].

The pathogenesis is not entirely clear. Aortic and mitral valves are the 2 most common sites of Lambl's filaments. It has been suggested that the high blood flow causes trauma to the heart valves, by inducing endocardial microfractures that occur at the valvular sites at the point of contact. Subsequently, fibrin is deposited, and there is a small formation of blood clots on the endothelial injury, with subsequent growth of the endothelium over a long period of time.

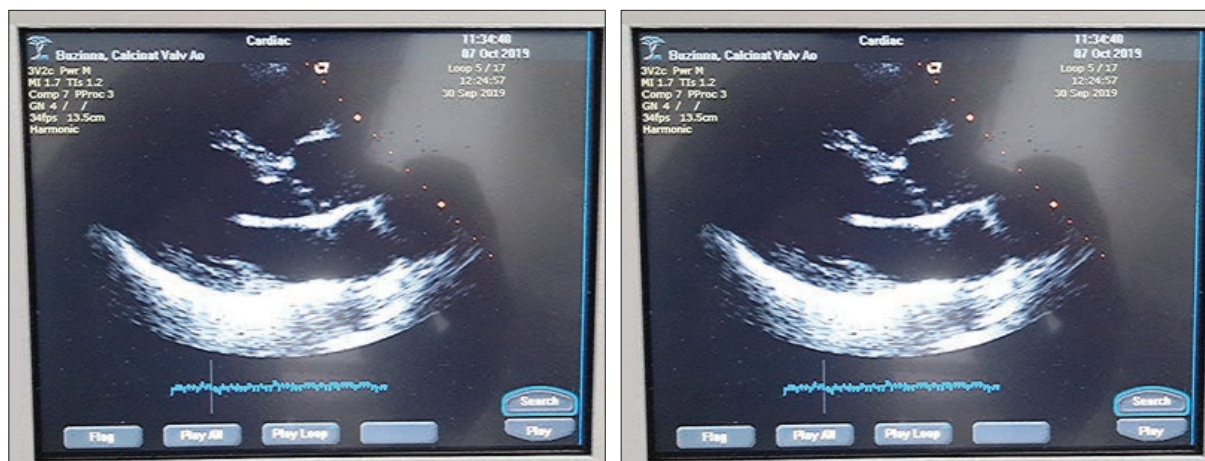
The pathogenesis of embolic events that cause distal trophic disturbances and trauma, such as ischemic stroke, acute coronary syndrome, or pulmonary embolism, remains unclear. While the possibility of Lambl's filaments coming off and obturating seems obvious, fragments of torn valve filaments are not usually observed in the arteries of the distal organs. There is a substantial evidence to suggest only a link between Lambl's growths and embolic events such as ischemic stroke and coronary syndrome. However, a direct causal relationship has not yet been established [5, 7].

In September 2019, patient B. complained to her chief physician about general weakness, decreased ability to work, cough, chest pain, tachycardia (up to 120 beats/min), and an increase in systolic blood pressure to 180 mm Hg. Art. This condition is associated with a cat bite and vaccination with rabies serum. In the anamnesis of the patient's life Lyme disease with circular erythema, complicated by polyneuropathy, joint syndrome; chronic pancreatitis, gallbladder hypokinesia, hypothyroidism.

The patient was tested for hormonal panel of the thyroid gland (levels of T3, TSH, and ATPO are normal, level T4 is reduced); MRI on pituitary adenoma (data on the presence of bulky tumors in the brain at the time of examination were not obtained); troponins and D-dimer are normal, CPK is increased (345 units/l), CPK MV is normal; package 38138 (detected *Borrelia burgdorferi*, IgG antibodies, IgM antibodies are absent) indicates a chronic course of tick-borne borreliosis; serology for heartworm disease is normal. The echocardiogram (EchoCG) on the aortic valve revealed a filamentous structure protruding into the left ventricular cavity, mitral valve prolapse, contractile function of heart is not impaired (Figure).

After the re-examination on several ultrasound – apartments, it was found that the patient has Lambl threads on the wings of the aortic valve.

Treatment. There is no clear evidence for the treatment of Lambl's growths. Various approaches, both pharmacological and surgical, in the treatment of Lambl's threads are described.



Echocardiogram on the aortic valve

Asymptomatic course is best monitored closely by follow-up echocardiography. When stroke is ruled out, transesophageal echocardiography should always be performed to identify the potential cause of the condition. These include carotid duplex ultrasound, hypercoagulated treatment, and a complete assessment of the ascending, transverse, and arched segments of aorta. If the treatment remains negative without any identifying cause, patients can be treated with antiplatelet agents such as aspirin and clopidogrel/dipyridamole and/or couadin anticoagulation. In cases of recurrent strokes associated with Lambl's growths, surgery may be necessary [8].

Differential diagnosis. Lambl's filaments should be differentiated from fibroelastoma, valvular vegetation, myxoma, cardiac thrombi, nonbacterial vegetation, intracardiac helminths, other heart tumors and metastases. All of these formations have different potential for embolization. Distinguishing Lambl's growths and nonbacterial vegetation or fibroelastomas can be quite a challenge. Others are quite well differentiated by echocardiography.

Lambl's filaments are histopathologically similar to cardiac papillary fibroelastomas. The difference is that papillary fibroelastomas are larger, usually located away from the valve closure line and covered with several layers of endothelial cells, whereas Lambl's growths are smaller, located on the line of valve closure and covered with a single endothelial layer. Fibroelastomas are usually found on mechanically less irritated parts of valves and other parts of endocardium, unlike Lambl's filaments. Thus, fibroelastomas are more voluminous, can be attached to the valve through a leg or pedestal, and their surfaces may contain several protrusions similar to fingers. Because fibroelastomas are larger than Lambl's growths, they are more prone to embolization. It is usually recommended to remove fibroelastoma surgically [9].

Myxoma of heart is a primary intracavitary histologically benign tumor, which is most often localized in the cavity of the left (75 %) or right (20 %) atrium, very rarely – in the ventricles of heart. The etiology of myxoma has not been elucidated. Tumor cells are thought to develop from embryonic mucoid tissue or endothelium, followed by myxomatous degeneration. Myxoma is a solitary, rarely multiple neoplasm of the round or oval shape that does not extend beyond the subendocardial layers of heart. The appearance of a tumor resembles a polyp or a bunch of grapes with a diameter of 5 mm to 8-12 cm with a coarse or fine-grained surface, its weight can

reach 250 g. The surface of the tumor is shiny, the capsule is clearly defined. 50 % of patients develop systemic embolic complications due to tumor fragmentation. Coronary artery embolism can lead to acute myocardial infarction. Surgical removal of myxoma is the only radical method of treatment, and delaying the operation should be considered a gross error.

Vegetative valves associated with endocarditis are usually irregular masses with different echogenicity relative to other cardiac structures. These vegetations can be attached to the valve surface at any site and are independent of valve motility, may have other pathognomonic echo data such as valve abscesses and dehiscence, a clinical history of infectious endocarditis, and valvular dysfunction. Vegetations in endocarditis are a mass of platelets, fibrin, colonies of microorganisms and pro-inflammatory cells. The main difference from Lambl's threads is that vegetations cause heart failure, which is absent in this patient [5].

Non-bacterial vegetation such as thrombotic vegetation Libman Sacks can be placed on any part of the valve cover. These are somewhat rounded and non-percussive lesions that lack independent mobility [5].

Lambl's filaments should be differentiated from intracardiac helminthiasis such as diphylariosis and angiostrongyloidosis, which are also important in the symptoms of angina and stroke, but these parasitic diseases are more prevalent in the right heart, in the thickness of the heart muscle [10].

Due to the lack of a clear evidence, it is difficult to exclude Lambl's filaments as a potential source of embolism in cryptogenic stroke.

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НИТКИ ЛАМБЛА ЯК ПРИЧИНА КАРДІОГЕННОЇ ЕМБОЛІЇ СУДИН ГОЛОВНОГО МОЗКУ ТА КОРОНАРНИХ АРТЕРІЙ

Розглянуто конкретний клінічний випадок і наведено дані літератури про рідкісну знахідку у порожнині серця — нитки Ламбла. Гістологічно це клітини з фіброеластичної тканини, покритої стромою та епітеліальним шаром. Вони не мають судинної оболонки, тому позбавлені грануляційної тканини. Нарости Ламбла виявляються як на власних, так і на протезованих серцевих клапанах. Дуже рідко нитки можна також побачити на інших ділянках, крім серцевих клапанів, наприклад на сосочкових м'язах, передсердних перегородках та хордах. Ці утворення можуть бути помічені на ехокардіограмі випадково або у зв'язку із серйозними ускладненнями, такими як емболічний інсульт або гострий коронарний синдром після їх відриву та поширення у віддалені органи. Нарости Ламбла виявляються у дорослих і у дітей, однак поширеність вище у перших і збільшується з віком. За результатами аналізу поширеність досягає піку в осіб віком від 61 до 70 років, а після цього зменшується через старіння серцевих клапанів, які все більше і більше кальцифікуються. Захворювання частіше зустрічається у чоловіків, ніж у жінок.

Ключові слова: нитки Ламбла, інсульт, стенокардія.