Adult Cor Triatriatum Sinistrum as an Unusual Cause of Embolic Stroke

Clinical Case Portal

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Abstract

Cor Triatriatum Sinistrum is a congenital anomaly in which the left atrium is divided into 2 parts by a fold of tissue, a membrane, or a fibromuscular band. Classically, the proximal (upper or superior) portion of the left atrium receives the venous blood, whereas the distal (lower or inferior) portion is in contact with the atrioventricular valve and contains the atrial appendage and the true atrial septum that bears the fossa ovalis. The anomaly is almost always diagnosed in early childhood. The hemodynamics of the Cor

Triatriatum are similar to that of mitral stenosis and the most common presenting symptoms in adults are dyspnea, hemoptysis and orthopnea as a result of the obstruction to blood flow.

Case Report

A 71 year old woman was referred to our echocardiography laboratory for evaluation of a potential cardiac source of embolus. The patient had suffered an embolic stroke in the territory supplied by the middle cerebral artery. She was admitted with dysarthria and aphasia. Prior to this event, she had no other brain infarction or neurological symptoms.

Relevant pathological findings at clinical examination were a nonfluent dysphasia and a faint holodiastolic cardiac murmur at the apex with little radiation. The woman was not dyspnoic and no abnormal respiratory sounds were heard at the chest evaluation.

Chest X-Ray showed no signs of heart failure or cardiac decompensation.

A cerebral computerized tomography (CT) on admittance revealed a vanishing low-density area in the outer convex brain surface, basal ganglia and the posterior and anterior internal capsules.

The patient had a history of mild systemic arterial hypertension as risk factor for arteriosclerosis, but no history of arrythmia or dyslipidemia. During 72h monitoring in the stroke unit she was continuously in sinus rhythm. Blood pressure was normal on admission and throughout hospitalization. The initial standard laboratory values were not remarkable. There was no evidence for activated protein C resistance, prothrombin mutation, lupus anticoagulant or hyperhomocystinemia, as indicators for thrombophilia. A doppler assessment of the carotid arteries showed no evidence of stenosis or plaques.

A complete transthoracic (TTE) and transoesophageal echo (TEE) study was performed

On 2D transthoracic echocardiography a membrane dividing the left atrium into 2 chambers was visualised as a linear, echogenic structure extending from the anterosuperior to the posterolateral wall of the left atrium (Fig.1). On M-Mode echocardiography a linear echo arising from behind the aortic root, in its anterior half, could be seen. The membrane showed pronounced movement during the cardiac cycle: anterior during diastole and posterior during systole. All cardiac chambers were normal in size. The mitral valve leaflets showed normal movement in all views. The other heart valves presented no abnormalities.
Doppler transthoracic echocardiography revealed no turbulences in the vicinity of the mitral valve or of the atrial membrane.
To precisely define the anatomy of the membrane, its relation to other structures, and the pulmonary venous drainage pattern, a transoesophageal echocardiography was also performed. The membrane was found to be attached laterally to the junction of the left upper pulmonic vein and left atrial appendage, dividing the left atrium into 2 chambers. The proximal chamber received blood from the pulmonary veins and the distal chamber contained the left atrial appendage and the mitral valve. One big fenestration connected the 2 chambers. The fenestration was situated in the proximity of the left atrial appendage. No other associated findings like a patent foramen ovale, an atrial septal defect or an anomalous pulmonary venous return were seen. Solely an increased mobility of the atrial septum could be noted, which was associated with an atrial septal aneurysm (ASA). (Fig.2) The study also revealed no atherosclerotic plaque in the aortic arch.
Inside the left atrium there was an increased amount of smoke as an indicator of increased blood echogenicity. The spontaneous echocardiographic contrast was extending from the mitral valve towards the membrane and the left atrial appendage with a higher density in the vicinity of the fenestration (Fig.3).
The woman was medicated with warfarin as anticoagulation therapy and was later referred to a

rehabilitation facility for people with neurological deficiencies. Her dysarthria and aphasia gradually improved over the course of the hospitalization and to this date she is doing well without any evidence of recurrence.

Discussion

We report the case of a 71 year old woman presenting with a cardioembolic stroke. In search of a potential cardiac embolus source, we accidentally found on echocardiography a membrane dividing the left atrium into two chambers. This finding was consistent with cor triatriatum sinistrum[1,2]. No other cardiac anomalies were seen. The patient had never had any symptoms of heart disease, nor evidence for a congenital disorder of coagulation, or a family history of cerebrovascular disease. She had only mild risk factors for a brain infarction (age, mild hypertension).

In this patient an embolic stroke was the first manifestation of cor triatriatum sinistrum at 71 years of age. This congenital heart disease is diagnosed in almost all cases in childhood and is usually incompatible with normal hemodynamics[3].

A possible explanation for the unusual late onset of symptoms could be the large fenestration of the membrane which allowed an almost normal blood flow towards the mitral valve. Nevertheless it can be assumed that directly around this opening the propagation of the blood influx has diminished in the course of time, resulting in hemodynamics as in mitral stenosis, with increased pressure inside the left atrium. This may be the explanation for increased echocardiographic contrast, which was seen on the transoesophageal study. None of the classical features associated with spontaneous echocardiographic contrast[4] was found in our case. The heart chambers were normal in size, the function of the left ventricle was not impaired, the mitral valve showed no stenosis or other dysfunction. In addition the patient had never had atrial fibrillation or other cardiac diseases.

Chimowitz and colleagues[5] describe the association between left atrial spontaneous echo contrast and stroke in patients with mitral stenosis. This condition leads to blood stagnation and increased risk for thrombus formation which, in this case, could have lead to embolisation from the proximal part of the left atrium and migration through the aortic arch to the carotid and middle cerebral artery.

The incidental finding of an atrial septal aneurysm appears to have no significance in this case report. According to Burger et al[6], the risk of cerebrovascular events or embolic strokes with incidental ASA is low.

As far as we know, there are very few case reports of cor triatriatum in adults presenting with an embolic stroke or arterial embolism, especially in an older age. A search through the on-line and professional literature revealed no more than 6 published cases of adult cor triatriatum associated with brain infarction. However, all these cases had additional other combined congenital heart abnormalities[7,8,9,10,11] and/or atrial fibrillation[7,8,9]. Nonetheless the association between cor triatriatum sinistrum and stroke might be fortuitous in a 71 year old hypertensive patient.

Conclusion

This case report underscores the diagnostic value of echocardiography in patients with neurological disease of presumed embolic origin. Although the diagnosis of Cor Triatriatum Sinistrum is commonly suspected on transthoracic study, transoesophageal echocardiography is the procedure of choice in recognizing and defining the subdividing membrane and its hemodynamic sequelar as in the present case.

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Fig. 1 :
Transthoracic echocardiogram

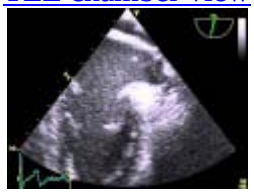
Fig. 2 :
Transoesophageal bicaval view showing a membrane arising from the fossa ovalis and dividing the left atrium in two parts.

Fig. 3 :

Transoesophageal view of the left ventricle, left atrial appendage and left atrium.

Video 1 :
Modified bicaval view

Video 2 :
TEE chamber view



Video 3 :
TEE-aortic and mitral valve long axis

Video 4 :
aortic valve short axis view