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Case Report

Renal infarction secondary to cor triatriatum sinister

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Abstract: A 30 year old, otherwise healthy man presented with flank pain and was ultimately found to have a right-sided renal infarction. Transthoracic echocardiography suggested, and then transesophageal echocardiography (TEE) confirmed, the presence of cor triatriatum sinister. Given the lack of other sources of emboli, this was felt to be the most likely source. We describe the case and both the echocardiographic and CT findings of this rare condition. This case demonstrates the need for TEE in some cases where 2D echocardiography is not sensitive enough to "rule out" cardio-embolic sources. This is only the second case in the literature of a systemic embolization due to cor triatriatum, and the first one in the Western literature.

Key words: Cor triatriatum, secondary renal infarction

Case Study

A 30 year old Caucasian man presented to the emergency room with complaints of severe flank pain on his right side lasting approximately two days. He had an unremarkable medical history and physical exam revealed only slightly elevated blood pressure. Computed tomography of the abdomen showed hypoperfusion of the lateral and superior portion of the right kidney, most consistent with infarct. A hypercoagulable workup was negative.

Due to the size of the infarction, and the lack of an alternative explanation, a transthoracic echocardiogram (TTE) was done to rule out an embolic source. The TTE revealed no significant abnormalities, but since there had been no adequate explanation of his infarction, a transesophageal echocardiogram (TEE) was ordered as further testing to rule out a cardiac embolic source.

The TEE showed normal left ventricular systolic function and no significant valvular stenosis or regurgitation. The left atrial appendage was free of clots and no aortic atherosclerosis was detected. A filamentous web of tissue was found in the left atrium. This web appeared to be attached to the atrial septum as well as the back wall of the left atrium. Computed tomographic coronary angiography, performed to better define the location and extent of the web, and confirmed this to be cor triatriatum sinister (**Figure 1**).

Given this patient's presentation with an embolic infarction and his previously asymptomatic state, he was started on anticoagulation therapy (warfarin) for prevention of recurrence of systemic embolization. Surgical resection was considered, but as he was otherwise asymptomatic, it was felt that anticoagulation alone would be preferable than open heart surgery and resection of the membrane. He has been followed closely in our clinic over the past two years and has remained event-free.

Discussion

Cor triatriatum sinister is a rare cardiac malformation that presents as a fibromuscular membrane separating the left atrium into two separate chambers. It accounts for approximately 0.1% of all reported congenital heart defects [1, 2]. The majority of reported cases of cor triatriatum occur in infants with symptoms of pulmonary venous obstruction[3].

Diagnosis in adults is most commonly the result

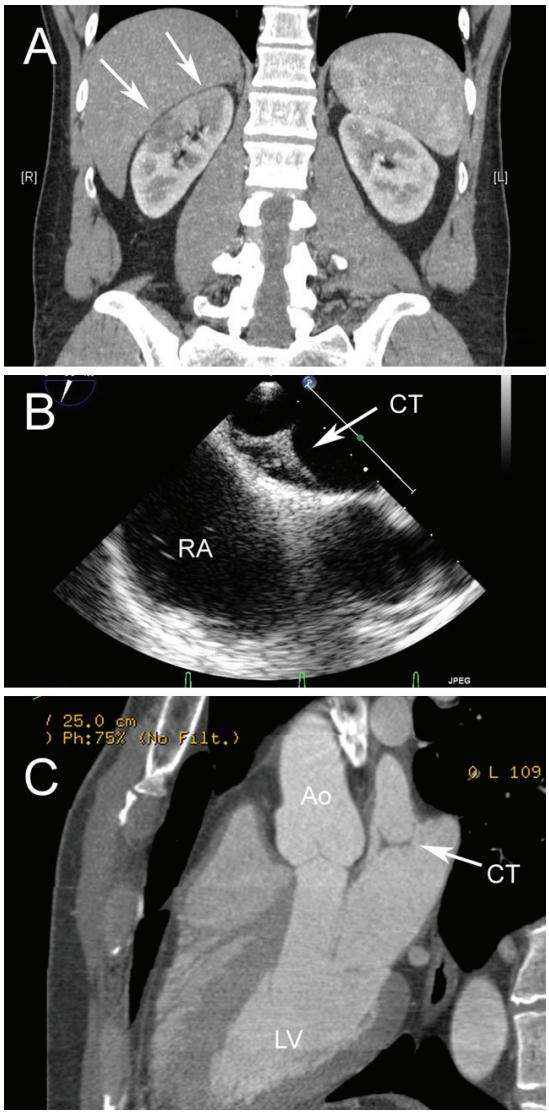

Figure 1. Computed tomographic coronary angiography. A) Coronal slices of an abdominal computed tomogram show right renal infarctions (arrows). B) Transesophageal echocardiogram showing a filamentous web of tissue (arrow) in the left atrium, with attachments on the atrial septum as well as the posterior atrial wall. C) Gate cardiac computed tomography demonstrating the location and attachment of the web of tissue in the left atrium. CT: cor triatriatum; Ao: aorta; RA: right atrium; LV: left ventricle.

of dyspnea and / or arrhythmias although other symptoms such as hemoptysis and orthopnea are sometimes seen [4-6]. Depending on the number and size of orifices perforating the membrane which divides the left atrium, cor triatriatum sinister may or may not present with symptoms, but when it does it often mimics mitral stenosis [7]. Onset of symptoms depends on the size of the orifice(s) separating the accessory atrial chamber from the true left atrial chamber, and it may be discovered as an incidental finding [8]. Additional cardiac abnormalities, such as mitral regurgitation and atrial septal defects, are associated with cor triatriatum [9].

Cases of cor triatriatum sinister have been initially misdiagnosed as mitral valve disease because of the way cor triatriatum mimics mitral stenosis clinically, radiographically, and electrocardiographically [10]. Diagnostic accuracy has been found to increase along with technological developments in the form of transthoracic and transesophageal echocardiography as well as cardiac CT [11].

The management of cor triatriatum sinister depends on the degree of obstruction between the two left atrial chambers. Surgical resection of the accessory membrane is recommended for patients presenting with obstruction, but no specific treatment is recommended for patients presenting with nonobstructive cor triatriatum [12]. Although surgical resection of the membrane has a very low rate of mortality, in the setting of an embolism, anticoagulation therapy is a conservative treatment for the prevention of future embolic events [1].

There have been only 7 previously reported cases of adult cor triatriatum discovered in the setting of an embolization. There has only been one case of systemic embolization including renal embolization, and none in the Western literature [13]. In 2009, Park, et al, compared the risk factors and treatments in 5 of these cases. Four of the 5 presented with stroke, with one patient presenting with a systemic infarction of multiple sites. Treatment of those patients included anticoagulation therapy, resection of the membrane, medical therapy (anticoagulation), and thrombectomy [14].

In conclusion, this report demonstrates the exceptionally rare finding of a cor triatriatum sinister in the setting of a systemic embolization. In patients without obvious cause for systemic embolization (especially young patients without a hypercoagulable state), this case illustrates the need for careful evaluation for structural abnormalities of the heart, as it can affect both

treatment and prognosis. In this situation, TEE and cardiac CT may improve the diagnostic yield over transthoracic echocardiography.

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