

Cor Triatriatum Sinistrum Presenting as Cardioembolic Stroke: An Unusual Cause of Adolescent Hemiparesis

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Cor triatriatum sinistrum is a rare congenital cardiac malformation, in which the left atrium (LA) is divided into two distinct chambers by a fibromuscular membrane. Classically, the proximal (upper or superior) chamber of the LA receives pulmonary venous connections, whereas the distal (lower or inferior) chamber contains LA appendage and true atrial septum containing fossa ovalis. The distal chamber is in continuity with the atrioventricular valve, while the two chambers communicate through a defect in the membrane. The hemodynamics of cor triatriatum sinistrum are similar to that of mitral stenosis due to obstructive property of membrane. The majority of reported cases of cor triatriatum occur in infants with symptoms of pulmonary venous obstruction, with adult cases being rare. Herein, we describe an unusual case of cor triatriatum in a 17-year-old boy who presented for the first time with embolic cerebral infarction with left hemiparesis. (Echocardiography 2014;31:E120–E123)

Key words: cor triatriatum

Case Summary:

A 17-year-old previously healthy male who was admitted in the department of neurology with sudden onset left hemiparesis and dysarthria was referred to the cardiology department for further investigation of suspected cardioembolic stroke. Computed tomography (CT) (GE Medical Systems, light speed, VCT, 64-slice, Waukesha, WI, USA) and magnetic resonance imaging (MRI) (Signa Excite 1.5 Tesla, GE Medical Systems) of brain had revealed infarction of right middle cerebral artery territory of brain (Fig. 1). He had uneventful medical history and had no significant risk factors for stroke, nor any congenital disorder of coagulation and no significant family history of cerebrovascular disease. There was no previous history of heart disease although he confirmed occasional shortness of breath on exertion with palpitations for last 1 year. Blood pressure was normal at the time of presentation and during the hospital stay. Positive findings on physical examination included an irregularly irregular pulse and a faint holodiastolic murmur at the cardiac apex. Electrocardiogram (ECG) (Sanborn Series, Philips Medical Systems, Andover, MA,

USA) showed atrial fibrillation with a fast ventricular rate. Chest radiograph showed a dilated atrial shadow. Laboratory data including risk factors for hypercoagulopathy (which included antithrombin III, protein C, protein S, and antiphospholipid antibodies) were not remarkable and bilateral carotid Doppler ultrasound was free of any atheromatous plaque.

A two-dimensional transthoracic echocardiography (TTE) (HD-11XE, Philips Ultrasound, Bothell, WA, USA) revealed a large left atrium (LA) which was divided into two by a membrane like echo dense structure (movie clip 1). This was attached medially to the interatrial septum (IAS) and laterally to the lateral wall of LA adjacent to the left atrial appendage (LAA), giving rise to a proximal small postero-superior chamber, and a distal large anterior-inferior chamber of LA, suggesting a cor triatriatum (Figs. 2 and 3). The membrane showed pronounced movement during the cardiac cycle (movie clip 2.). The transesophageal echocardiography (TEE) (HD-11XE) was done to compliment TTE findings (Fig. 4) (movie clip 3) and to search for embolic source. On TEE, thick spontaneous echo contrast (SEC) was seen in the distal chamber and a soft clot was seen in LAA, which was presumed to be the cardioembolic source (Fig. 5). There was no significant pressure gradient observed within the LA using color flow Doppler imaging. The IAS was

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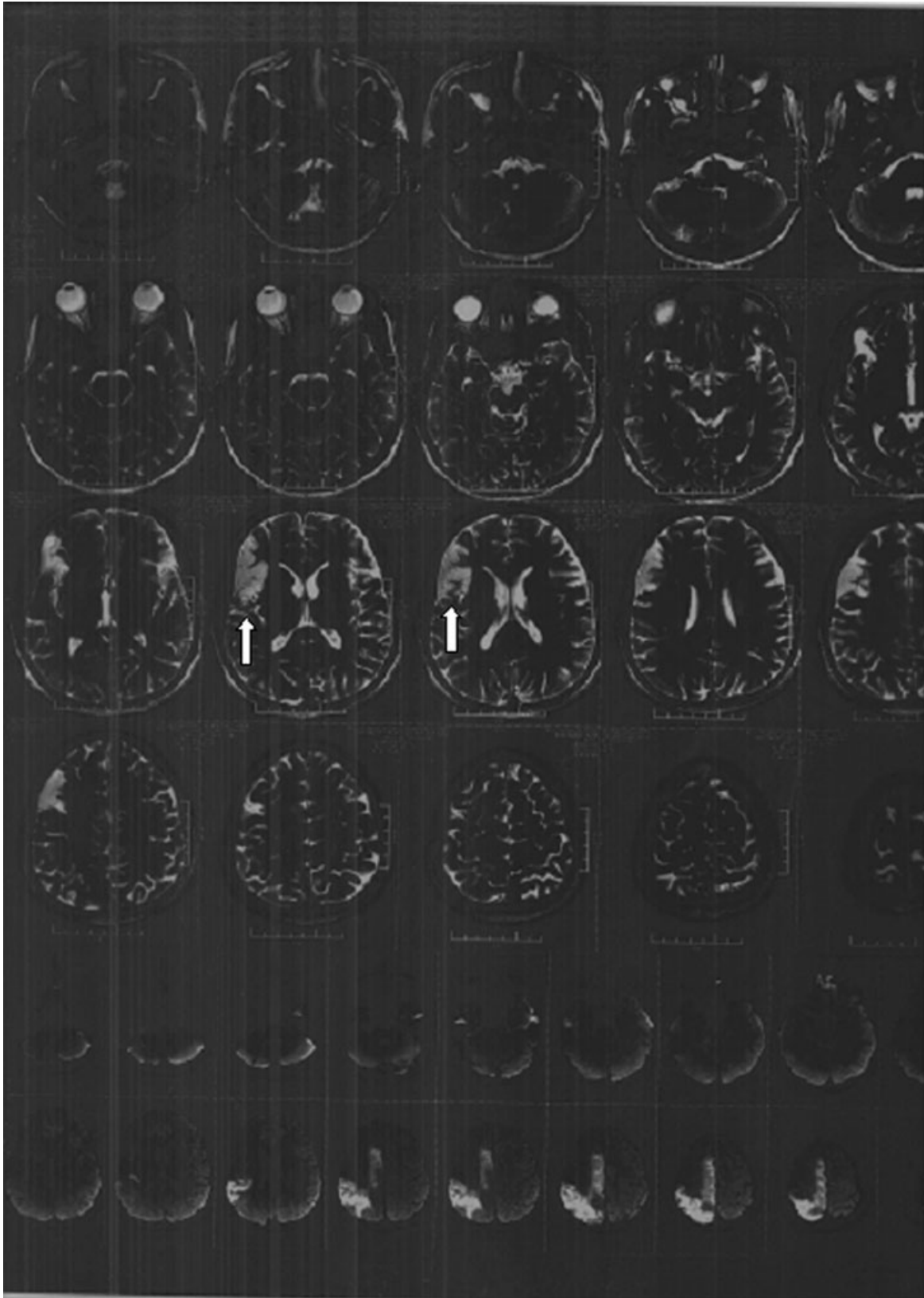


Figure 1. Diffusion-weighted magnetic resonance imaging of the brain shows high signal intensity in the right cerebral hemisphere suggestive of an acute ischemic infarction in the territory of the right middle cerebral artery (arrows).

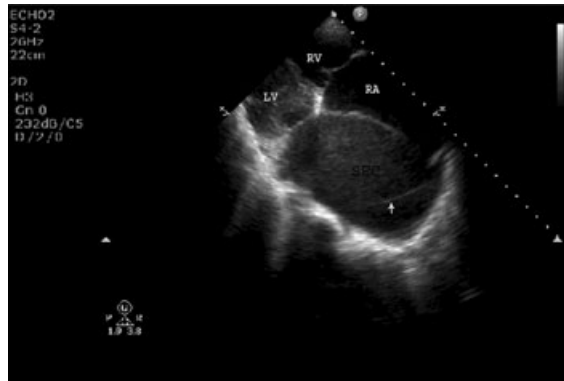


Figure 2. Transthoracic modified four-chamber (4C) echocardiographic view showing membrane (arrow) crossing left atrium (LA) and dividing it into upper and lower chamber. Note lower chamber containing left atrial appendage (LAA) is full of spontaneous echo contrast (SEC). Left ventricle appears obliterated due to modified 4C view.

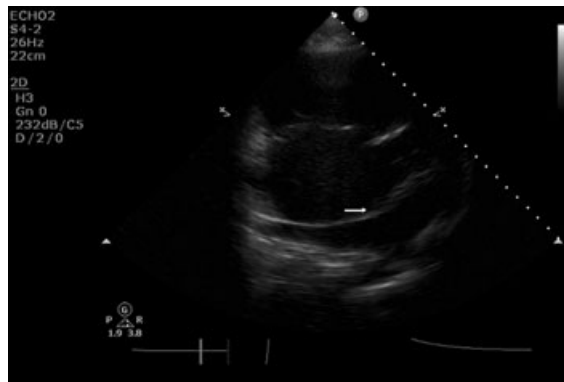


Figure 3. Another transthoracic modified echocardiographic view showing fibromuscular membrane (arrow) crossing left atrium (LA).

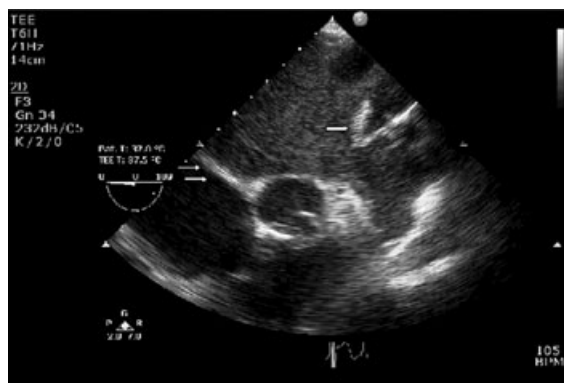


Figure 4. Transesophageal echocardiographic (TEE) view showing fibromuscular membrane attached to left atrial appendage (LAA) (arrow). Interatrial septum (IAS) is seen on left (double arrow).

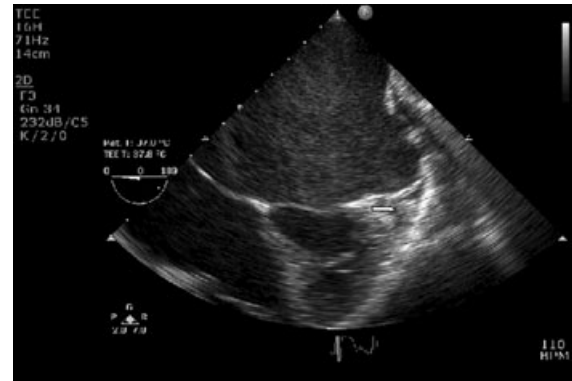


Figure 5. Modified transesophageal echocardiographic (TEE) view shows left atrial appendage containing clot (arrow).

intact and all pulmonary veins drained into the proximal superior chamber. The distal inferior chamber, which contained the LAA, communicated with the left ventricle through a morphologically normal mitral valve. There was no other associated abnormality seen on echocardiography.

The final diagnosis rendered in the adolescent was embolic stroke associated with cor triatriatum sinister, atrial fibrillation, and LAA clot. The patient was managed conservatively with anticoagulation therapy which was tolerated well. His hemiparesis and dysarthria gradually improved and to date he has not shown any evidence of recurrence.

Discussion:

Cor triatriatum is a rare congenital defect first reported by Church in 1868.¹ It is found in only 0.1% of cases of congenital heart diseases.² Cor triatriatum sinister results from the persistence of a fibromuscular membrane in the LA, dividing it into a proximal posterosuperior chamber receiving pulmonary veins and a distal anteroinferior chamber containing the LAA and communicating with the mitral orifice. Embryologically, this condition is thought to arise from the failure of inclusion of the pulmonary veins into the left posterior atrial wall.

Cor triatriatum dextrum is extremely rare and results from the complete persistence of the right sinus valve of the embryonic heart. The membrane divides the right atrium into upper and lower chambers. The upper chamber receives the venous blood from both vena cavae and the lower chamber is in contact with the tricuspid valve and the right atrial appendage.

The majority of reported cases of cor triatriatum sinister occur in infants with symptoms of pulmo-

nary venous obstruction. Although, It is frequently an isolated finding in adults,^{3,4} cor triatriatum has been reported in association with ostium secundum atrial septal defect (ASD), dilated coronary sinus due to persistent left superior vena cava, and bicuspid aortic valve in adults.⁵ Diagnosis in adults is most commonly the result of dyspnea or arrhythmias and often it mimics mitral stenosis.⁶ The natural history of cor triatriatum depends on the effective size of the orifice(s) in the diaphragm and the presence and location of the ASD. When small, it usually presents in infancy with reduced cardiac output, pulmonary venous hypertension, and cardiac failure. In patients with large communications, the diagnosis may present only in adulthood with development of fibrosis and calcification in the orifice of the separating membrane, along with development of mitral regurgitation and atrial fibrillation.³ The embolic presentation is extremely rare both in children and adults.

The management of cor triatriatum sinister depends on the degree of obstruction between the two LA 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.⁷ In the setting of an embolism, anticoagulation therapy is a conservative treatment for the prevention of future embolic events.⁸ A search of existing literature revealed only 9 previously reported cases of adult cor triatriatum discovered in the setting of embolism and no more than 7 published cases of adult cor triatriatum associated with cerebral infarction⁹ and none in an adolescent. In 2009, Park, et al.¹⁰ 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.¹⁰

Conclusion:

In conclusion, this report demonstrates the exceptionally rare finding of a cor triatriatum sinister in an adolescent presenting as embolic stroke with hemiparesis, underscoring the impor-

tance of careful evaluation for structural abnormalities of the heart in patients with systemic embolization. The natural history and complications of cor triatriatum sinister have been poorly documented, which relates primarily to the small numbers available for review. Our work adds further to the published literature available on this rare congenital anomaly.

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie clip S1. Modified TTE 4C view.

Movie clip S2. Modified TTE 4C view with anterior angulation. Pronounced movement of fibromuscular membrane is seen. Dense SEC seen in anterior-inferior chamber.

Movie clip S3. Modified TEE view showing fibromuscular membrane, SEC in LA and soft clot in LAA.