Adult Cor Triatriatum Presenting as Cardioembolic Stroke

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Abstract

Cor triatriatum is a rare congenital cardiac malformation characterized by a fibromuscular membrane that divides the left atrium into two distinct chambers. In almost all cases, it is diagnosed in childhood, whereas adult cases are extremely rare. Herein, we describe an unusual case of cor triatriatum in a 55-year-old woman who presented with embolic cerebral infarction. The patient experienced sudden-onset, transient left-sided homonymous hemianopsia and echocardiography and multidetector computed tomography detected a membrane-like structure across the left atrium, confirming the diagnosis of a cor triatriatum. The laboratory examination for hypercoagulopathy was negative. She was conservatively treated with anticoagulation and her neurological manifestation gradually improved.

Key words: cor triatriatum, cerebral infarct, congenital heart disease, embolism


Introduction

Cor triatriatum is a rare congenital cardiac malformation in which a fibromuscular membrane divides the left atrium (LA) into two distinct chambers (1). The hemodynamics of the cor triatriatum is similar to that of mitral stenosis and the most common presenting symptom of cor triatriatum in adults is dyspnea, hemoptysis, and orthopnea as a result of the obstructive function of the intra-atrial membrane (2). In almost all cases, it is diagnosed in childhood, whereas adult cases are very rare (3-5). In this report, we present an unusual case of cor triatriatum in a 55-year-old woman initially presenting with embolic cerebral infarction in whom the diagnosis was delayed until adulthood.

Case Report

A 55-year-old previously healthy woman who had visited the department of neurology outpatient clinic of our institution due to sudden-onset, transient left-sided homonymous hemianopsia was referred to this cardiovascular center for further investigation of source for cardioembolic stroke. Previously, she had uneventful medical history and had no classical risk factors for stroke/embolism such as hypertension, diabetes mellitus, dyslipidemia, and congenital disorders of the coagulation system and there was no significant family history for cerebrovascular disease. There was no previous history of heart disease and she denied any cardiac symptoms such as dyspnea or orthopnea. On physical examination, her pulse rate and blood pressure were normal. Neurologically, a left-sided homonymous hemianopsia was present. Cardiac examination revealed regular heart rate and rhythm with normal heart sounds and no murmurs. Chest radiography showed a normal-sized heart with normal pulmonary vascularity. The electrocardiogram showed normal sinus rhythm. Her initial laboratory data were not remarkable. Diffusion-weighted magnetic resonance imaging of the brain revealed acute ischemic infarction of the right occipital lobe (Fig. 1). Magnetic resonance angiography of the intracranial and extracranial arteries and carotid doppler ultrasound were free of atheromatous plaque. A presumptive diagnosis of
embolic stroke was made. A two-dimensional transthoracic echocardiography and transesophageal echocardiography (TEE) revealed a membrane-like, echo-dense structure attached to the interatrial septum medially and to the left atrial appendage laterally, which divided the LA into posterior-superior and anterior-inferior chambers suggesting a cor triatriatum (Fig. 2). There were no apparent pressure gradients within the LA observed using color-flow doppler imaging. Although there was no definite evidence for embolic source such as thrombus in LA appendage, a mild spontaneous echo contrast (SEC) confined to the dilated proximal chamber of LA was observed and it was presumed to be her cardioembolic source. TEE also revealed no atherosclerotic plaque in the aortic arch. The interatrial septum was intact. ECG-gated 64-slice multidetector computed tomography (MD CT) subsequently performed for the evaluation of combined cardiac anomaly also clearly showed the distinct intra-atrial septum in LA, which divided LA into an proximal accessory chamber and a distal true chamber (Fig. 3). All pulmonary veins drained correctly into the accessory LA and the true LA gave rise to the left atrial appendage and communicated with the mitral valve. The diameter of communicating large openings in the membrane was determined to be 1 centimeter. Risk factors for hypercoagulopathy including homocysteine, antithrombin III, protein S, protein C and antiphospholipid antibodies were all normal or negative. Finally, she was diagnosed with embolic stroke associated with cor triatriatum and conservatively treated with anticoagulation therapy. Her neurological manifestation gradually improved and to date she is doing well without any evidence of recurrence.

**Discussion**

Here, we describe a case of a 55-year-old woman with cor triatriatum initially presenting with cardioembolic brain infarction. This patient had a membrane in the LA consistent with cor triatriatum that partially divided the LA into two chambers and no other combined congenital heart disease. The onset of symptoms and clinical manifestations depends on the size and number of the defects within the separating membrane, the morphology of the defects, and the presence of other cardiac anomalies (2). In the present case, the patient remained asymptomatic until 55 years of age and the first manifestation was embolic stroke. To our knowledge, there are only a few case reports of adult cor triatriatum presenting as embolic stroke or arterial embolism (6-10). A MEDLINE/PUBMED literature search was performed (key word: cor triatriatum stroke embolism) to identify all similar cases in the medical literature and five cases of adult cor triatriatum presenting as embolic stroke (four cases) or arterial embolism (one case) were available from the literature review. A review of the literature on embolic events in adults with cor triatriatum is summarized in Table 1. The mean age at diagnosis was 49 years (30-65 years) with the median age of 55 years, which was the same age as the present patient. The woman to man ratio was 3:2. Brain imaging in patients with embolic stroke (four cases) revealed middle cerebral artery (MCA) territory infarction (one case), brain swelling (one case) and multiple cerebral infarctions (one case). Regarding the clinical stroke risk factors, there were atrial fibrillation (three cases), diabetes mellitus (three cases), hypertension (three cases) and hyperlipidemia (two cases). The echocardiographic features related with cardioembolic stroke include left atrial enlargement (three cases), type 2 atrial septal defect (one case), mitral regurgitation (one case), left atrial thrombi (two cases) and spontaneous echo contrast in accessory atrium (one case). All of the previous cases had the features strongly suggestive of cardioembolic stroke such as intracardiac thrombus...
Figure 3. Cardiac multidetector computed tomography (upper panels, sagittal view and lower panels, five-chamber view) reveals an intra-atrial membrane (arrowheads) in the left atrium with large communicating openings (arrows). LA: left atrium, aLA: accessory left atrium

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Af: atrial fibrillation; HTN: hypertension; DM: diabetes mellitus; Ct: Cor triatriatum; LA: left atrium; LAE: left atrial enlargement; NR: not reported; ASD: atrial septal defect; SEC: spontaneous echo contrast; MCA: middle cerebral artery; RCA: right coronary artery; RM: resection of membrane of cor triatriatum

(cases 1 and 2), multiple cerebral infarctions (case 3), systemic thromboembolism (case 2 and case 5) and young-aged patient and combined atrial septal defect (case 4). Compared with these previous case reports, the present case did not
have definite risk factors for cardioembolic stroke such as atrial fibrillation and we were not able to detect any intracardiac thrombus that would be the source of cerebral embolism. And, the patient did not have multiple cerebral infarctions on brain imaging either peripheral artery embolism. However, she had the features suggestive of cardioembolic stroke such as no significant atherosclerosis on cerebral vessels and aortic arch and sudden-onset hemianopsia due to PCA territory infarction. In our case, the intra-atrial communication was wide and there was no significant transmembrane flow obstruction between the proximal and distal chambers. Although no definite intracardiac thrombus was demonstrated and the embolic source in this case was obscure, blood stagnation in the proximal chamber which manifests as spontaneous echo contrast may predispose intracardiac thrombus formation. The embolic cerebral infarct most likely developed due to the blood clots which embozilized from the proximal part of the LA and lodged in the most likely developed due to the blood clots which embozilized from the proximal part of the LA and lodged in the occipital lobe via the posterior cerebral artery. Although not documented in our patient, paroxysmal atrial fibrillation also occurred, blood stagnation in the proximal chamber which manifests as spontaneous echo contrast may predispose intracardiac thrombus formation. The embolic cerebral infarct most likely developed due to the blood clots which embolized from the proximal part of the LA and lodged in the occipital lobe via the posterior cerebral artery. Although not documented in our patient, paroxysmal atrial fibrillation also may be another potential explanation for the cause of cardioembolic stroke in this patient.

The value of echocardiography in patients with neurological disease of presumed embolic origin is well established (11). For cor triatriatum, echocardiography is the most commonly used imaging technique for the diagnosis and careful echocardiography could be the procedure of choice in identifying the subdividing membrane as in the present case (12, 13). Although the diagnosis is commonly suspected on transthoracic study, transesophageal echocardiography (TEE) is frequently needed to precisely define the anatomy of the membrane, and the degree of obstruction caused by this structure (13-15). The cardiac MDCT, a new and powerful non-invasive imaging modality, provides a comprehensive anatomic evaluation of cor triatriatum (15). It can help the clinician to see a distinct septum in the LA, to evaluate the size and number of fenestrations, and to identify associated congenital cardiovascular anomalies (16).

Management of cor triatriatum depends on the degree of obstruction between the LA chambers (17). Surgical resection of the accessory membrane is indicated in symptomatic patients with obstructive cor triatriatum (17). The present patient had no signs of pulmonary hypertension, and remained asymptomatic. Also, she did not have atrial fibrillation or other combined intracardiac defect. Therefore, although surgical correction is relatively simple with low mortality (18), she was conservatively treated with anticoagulation for prevention of recurrence of embolism. In conclusion, this case report underscores the importance of being aware of possible embolic complications in order to avoid such rare complications even in asymptomatic adults with cor triatriatum.

References


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