A Rare Association of Pulmonary Thromboembolism in Non-obstructive CorTriatriatum

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Abstract

This is to report incidently detected non obstructive cortriatriatum in an lady who presented with acute pulmonary thromboembolism and its clinicoradiological association. Case is reported due to its rarity.

Keywords: Cortriatriatum, Acute pulmonary thromboembolism, Trans esophageal echocardiography, CT pulmonary angiography.

A 43 year old lady presented to the emergency department with acute worsening of pre-existing dyspnea. She was undergoing treatment for cerebrovascular accident, hypothyroidism and paranoid schizophrenia from a nearby hospital for past few years.

She had pallor, was tachypneic, had tachycardia and blood pressure of 110/70 mmHg. She also had loud palpable P2 with narrow splitting of second sound. Other systems examined were within normal limits excluding her previous debility.

Investigations: ECG revealed sinus tachycardia with QRS axis of +90 degrees and S1Q3T3 pattern. Chest radiogram revealed clear lung fields with prominent pulmonary artery and trans-thoracic echocardiography showed RA/RV dilatation with systolic RV dysfunction, and raised pulmonary artery pressures (RVSP>64mmHg) (figure1A).

Figure 1

Figure1A: 2D echocardiography showing Cortriatriatum membrane, labelled as ‘a’

Plasma D-Dimer was elevated (>10000 ng/ml). All these findings strongly suggested the diagnosis of

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acute (possibly on chronic) pulmonary thromboembolism. Further descriptive analysis of 2D echocardiography and colour Doppler study also revealed incidentally the presence of a membrane separating left atrium without obstructing the flow through the mitral valve (figure 1B).

**Management**

She was thrombolysed with Tenecteplase. Serial 2D echo after thrombolysis demonstrated gradually decreasing pulmonary artery pressures with non obstructive cortedriatium. Serial CT Pulmonary angiography showed improved blood flow towards right pulmonary artery. Patient was discharged on warfarin and other symptomatic drugs. She was conservatively managed. Since it was a non-obstructive type cortedriatum, no surgical intervention was required.

**Discussion**

Cortedriatum is a rare cardiac malformation that accounts for approximately 0.1% of all reported congenital heart defects.\(^1\)\(^,\)\(^2\) In this malformation, failure of resorption of the common pulmonary vein results in a left atrium divided by an abnormal fibromuscular diaphragm into a posterosuperior chamber receiving the pulmonary veins and ananteroinferior chamber giving rise to the left atrial appendage and leading to the mitral orifice.\(^3\) The communication between the divided atrial chambers may be large, small, or absent, depending on the size of the opening(s) in the diaphragm, which determines the degree of obstruction to pulmonary venous return. The majority of reported cases of cortedriatum occur in infants with symptoms of pulmonary venous obstruction.\(^4\) Obstructed cortedriatum in adults most often present with dyspnea and / or arrhythmias although other symptoms such as hemoptysis and orthopnea are sometimes seen.\(^5\)\(^,\)\(^6\) Onset of symptoms depends on

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**Figure 1B**: Colour Doppler 2D image demonstrating non-obstructive membrane in left atrium

**Figure 3**: CT Pulmonary angiography with features suggesting pulmonary thromboembolism

So for identifying the source of embolus and correct description of membrane patient was taken up for Transesophageal echo (TEE) which revealed similar findings as 2D echo and without any source of embolism (Figure 2).

She was worked up for thrombophilic states, which yielded a positive Anti nuclear antibody but tests for specific antibodies were negative. Thus immunological data were inconclusive of any specific etiology that could explain the source of embolus.
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. Cortriatriatum with severe pulmonary hypertension complicated by chronic pulmonary artery thrombosis is very rare. Elevations of both pulmonary venous pressure and pulmonary vascular resistance may result in severe pulmonary artery hypertension. Depending on the number and size of orifices perforating the membrane which divides the left atrium, cortriatriatum sinister may or may not present with symptoms, but when it does it often mimics mitral stenosis.

Conclusion

This case report demonstrates the exceptionally rare finding of a cortriatriatum sinister in the setting of a pulmonary embolization. In patients without obvious cause for pulmonary embolization (especially young patients without a hypercoagulable state), there should be a careful evaluation for structural abnormalities of the heart. In this situation, TEE and CT Angiography may improve the diagnostic yield over transthoracic echocardiography.

References