Pseudo-cor triatriatum dexter: Rare and asymptomatic but clinical consequences are contemplative

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ABSTRACT

Introduction: Congenital anatomic variants are more frequent in the right atrium. Complete or incomplete bisection of the right atrium by a membrane-like fold across the cavity is designated as cor triatriatum dexter. Even though most of the cases of congenital atrial remnants are asymptomatic, the clinical consequences in the presence of cardiovascular diseases are very contemplative.

Case Report: We report a case of an elderly patient with acute coronary syndrome and an incidental finding of pseudo-cor triatriatum dexter. Coronary angiography and intervention was done without any complications.

Conclusion: Being an extremely rare entity, a partial form of cor triatriatum dexter is an important congenital anomaly to document on echocardiography, and clinical implications could be correlated by clinicians during the management of multidisciplinary diseases.

Keywords: Acute coronary syndrome, Cardiac catheterization, Pseudo-cor triatriatum

INTRODUCTION

Cor triatriatum is one of the rare forms of acyanotic congenital heart disease (ACHD) with an estimated reported incidence rate of 0.1% of all congenital cardiac anomalies [1]. The left atrial form is known as cor triatriatum sinistrum (CTS) and the right atrial form is known as cor triatriatum dexter (CTD). Depending upon the extent of membrane dissection of the atrial cavity, it is either complete or partial in anatomy. Partial forms of cor triatriatum are usually asymptomatic unless associated with other anomalies like atrial septal defect (ASD), right-sided abnormalities like hypoplasia or atresia of the tricuspid valve or pulmonary trunk, or obstruction of right ventricular inflow tract. We report a case of an unidentified partial form of CTD in acute coronary syndrome with potential clinical implications.

CASE REPORT

A 74-year-old man presented to the emergency department with the acute coronary syndrome as non-ST elevation myocardial infarction and acute pulmonary edema. Detailed echocardiography was done for ejection fraction and hemodynamic evaluation. The findings include bi-atrial enlargement, normal interatrial septum (IAS), mild tricuspid regurgitation (TR), moderate pulmonary hypertension (PH) by TR jet method, non-dilated pulmonary trunk, left ventricular ejection fraction – 44%, and severe interventricular septum (IVS) hypertrophy prominently at the basal end. Interestingly, the right atrium (RA) was found double-chambered by a membranous echogenicity. A bright non-mobile echogenicity, equating like a membrane, was visualized with attachments at one end at the mid-section of the IAS and the other end at the posterior wall of RA as shown in the four-chamber view (Figure 1A) and the subcostal view (Figure 1B). The color
Eustachian valve, Chiari network, conditions are the absence of any transmembrane gradient or associated RA with end attachment to the posterior wall and IAS. In immobile membranous structure is linear and bisecting pulmonary embolism [3]. As in the presented case, the forms a potential nidus for thrombus formation and in the RA cavity. This fenestrated mesh is mobile and the posterior wall of LA as a reticular curvilinear network of the right valve of sinus venosus and it progresses over Chiari network is also a remnant of incomplete resorption to RA, the possibility of the Eustachian valve is ruled out. The membrane is arising 2.485 mm distal to IVC junction protrudes into the atrial cavity. As per findings of TTE, it arises from the superior rim of IVC and another end foramen ovale during fetal circulation [2]. Therefore, the inferior vena cava (IVC) to the left atrium (LA) via pulmonary circulation and direct oxygenated blood from and CTD.

**DISCUSSION**

The right atrial membranous structures are subject to a variety of anatomic variants. The congenital remnant stems from incomplete embryological regression of sinus venosus. There is large variability in dimension, density, and configuration of this remnant. The extent of a persistent remnant is visible as ridge-like echogenicity on transthoracic (TTE) and transesophageal (TEE) echocardiography. The differential diagnosis for this case includes a prominent Eustachian valve, Chiari network, and CTD.

The Eustachian valve is a functional valve to bypass pulmonary circulation and direct oxygenated blood from the inferior vena cava (IVC) to the left atrium (LA) via foramen ovale during fetal circulation [2]. Therefore, it arises from the superior rim of IVC and another end protrudes into the atrial cavity. As per findings of TTE, the membrane is arising 2.485 mm distal to IVC junction to RA, the possibility of the Eustachian valve is ruled out. Chiari network is also a remnant of incomplete resorption of the right valve of sinus venosus and it progresses over the posterior wall of LA as a reticular curvilinear network in the RA cavity. This fenestrated mesh is mobile and forms a potential nidus for thrombus formation and pulmonary embolism [3]. As in the presented case, the immobile membranous structure is linear and bisecting RA with end attachment to the posterior wall and IAS. In the absence of any transmembrane gradient or associated anomalies, it is described as pseudo-cor triatriatum dexter (pCTD). A cardiac computed tomography scan can easily identify the atrial cavity membrane or fibrous ridge with a three-dimensional volume-rendered image analysis [4]. Also, cine cardiac magnetic resonance imaging (CMR) can identify the flow-related abnormalities of the membrane as a low-intensity signal flow from fenestrations in contrast to high-intensity blood flow [5].

The symptomatology and clinical outcome is solely dependent upon the mechanical restriction of intra-atrial flow and right ventricular inflow. Our case report is asymptomatic and is an incidental presentation of pCTD. However, differential diagnosis is very important for the prevention of progression of any added risk factors for this patient. The clinical significance of pCTD is also important as this patient underwent interventional treatment for ischemic heart disease. In case of any additional interventional treatment like temporary or permanent pacemaker implantation, the pacing wire may face resistance from the membrane. Any rupture of the membrane can also provoke fragment emboli or intra-atrial clot formation. In the case of septicemia, this membrane may serve as a nidus for vegetations. Possibly, any fibrosis or calcification of this anomalous tissue can restrict the diastolic function of the RA and may provoke congestive symptoms simulating increased preload.

**CONCLUSION**

Our case report reveals that even the asymptomatic forms of congenital heart diseases can be of significant clinical importance when comorbid conditions are managed. Even though a pseudo-form of CTD is not repaired in asymptomatic cases, the complications like stasis or clot formation, right-heart interventional treatment, extra-valvular intra-cardiac vegetation foci, and age-related fibrosis or calcification of the membrane should not be missed and a high level of suspicion should be maintained. Being an extremely rare entity, a partial form of CTD is an important congenital anomaly to document on echocardiography, and clinical implications can be correlated by clinicians during the management of multidisciplinary diseases.

**LEARNING OBJECTIVE**

1. Pseudo-cor triatriatum dexter is a rare congenital abnormality with a variety of morphological presentations.
2. Presence of ischemic heart disease in pseudo-cor triatriatum dexter poses unique treatment challenges.
3. Identification of pseudo-forms of cor triatriatum on transthoracic echocardiography is critical for individual risk stratification.
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Author Contributions

Keyur Vora – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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