SCIE DOI: 10.14744/scie.2017.79926
South. Clin. Ist. Euras. 2017;28(1):61-63

Case Report

A Rare Case: Cor Triatriatum Dexter

Ali Orgun, Alper Hazım Gürsu, Emine Azak, Ayşe Esin Kibar, İbrahim İlker Çetin

Department of Pediatric Cardiology, Ankara Children's Hematology Oncology Training and Research Hospital, Ankara, Turkey

Submitted: 02.11.2016
Accepted: 02.02.2017

Correspondence: Ali Orgun, Ankara Çocuk Sağlığı ve Hastalıkları Hematoloji Onkoloji Eğitim ve Araştırma Hastanesi, Çocuk Kardiyoloji Kliniği, 06110, Ankara, Turkey
E-mail: draliorgun@hotmail.com

Keywords: Cor triatriatum; echocardiography; rheumatic fever; three-dimensional.

ABSTRACT

Cor triatriatum dexter (CTD) is a rare congenital heart disorder that is result of persistence of the right valve of the embryonic sinus venosus. Clinical significance of CTD is related to degree of obstruction. Asymptomatic patients may be diagnosed during echocardiographic examination performed for unrelated reasons. Presently described is case of a 12-year-old girl with initial diagnosis of acute rheumatic fever whose echocardiogram revealed presence of CTD. Aim of this report was to emphasize that diagnosis of CTD can be difficult and requires careful evaluation of the right atrium by echocardiogram.

INTRODUCTION

Cor triatriatum is a very rarely seen congenital heart disease. Its incidence among all congenital heart diseases is only 0.1%.[1] Most cases are known as classic cor triatriatum, namely cor triatriatum sinister (CTS). Cor triatriatum dexter (CTD), in which fibromuscular band is seen in the right atrium, is very rarely encountered, and most cases lead an asymptomatic course. Asymptomatic cases of CTD may be diagnosed during echocardiographic examination performed for other indications. Presently described is case of CTD detected during echocardiographic examination performed in our clinic for patient with indication of acute rheumatic fever (ARF). Diagnosis of rarely seen CTD is very difficult to make, and right atrium should be examined very carefully during echocardiographic procedure to establish diagnosis.

CASE REPORT

A 12-year-old female patient presented at outpatient clinic with complaints of swelling and painful right ankle persisting for 1 week. Personal history was unremarkable. Family history revealed that her father suffered from ARF. Some notable laboratory parameters were as follows: erythrocyte sedimentation rate, 91 mm/h; C-reactive protein, 13.9 mg/dL; white blood cell count, 16.900/mm^3; and antistreptolysin O, 759 IU. On physical examination, apart from auscultation of 1/6 early systolic murmur over mesocardiac focus and painful, swollen, and erythematous right ankle, no abnormal sign was found. Electrocardiographic examination results and peripheral oxygen saturation were within normal limits. Transthoracic echocardiograms demonstrated first-degree mitral insufficiency, and left ventricular ejection fraction was estimated at 67%. On 3-dimensional echocardiogram, fibromuscular membrane that was extension of crista terminalis in the right atrium localized in vicinity of junction between vena cava superior (VCS) and the right atrium was seen (Figure 1). Continuous-wave Doppler ultrasound (DUSG) images were obtained through opening in middle of the fibromuscular membrane. DUSG revealed presence of turbulent flow at rate of 1.5 m/s with no obstructive lesion (Figure 2). Based

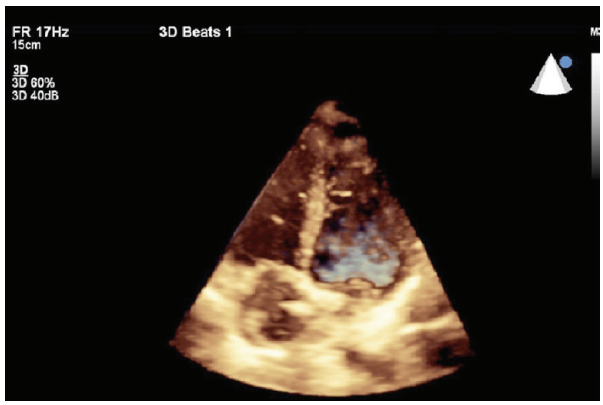

Figure 1. Appearance of the fibromuscular membrane on 3-dimensional echocardiogram.

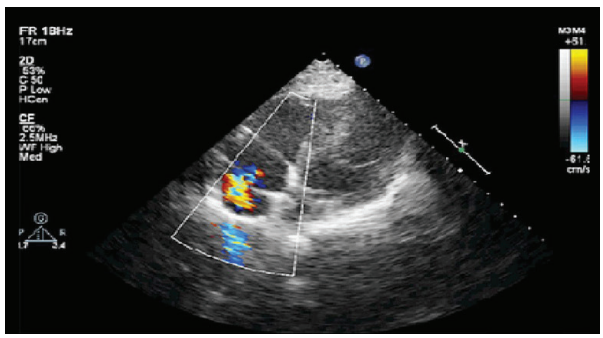

Figure 2. Color Doppler image of turbulence seen in the fibromuscular membrane.

on these findings, the patient received diagnoses of ARF carditis and CTD. She was hospitalized with indication of ARF carditis, and prednisolone treatment at daily dose of 2 mg/kg was initiated. During follow-up, prednisolone dose was decreased, and naproxen (15 mg/kg/d) was added to existing treatment. Follow-up protocol on ambulatory basis was planned with indication of CTD. The patient is still under ambulatory follow-up treatment with diagnoses of ARF and CTD. Informed consent was taken from the patient's family.

DISCUSSION

Cor triatriatum is very rarely seen, and most cases are CTS, presence of fibromuscular membrane in the left atrium. CTD is very rarely seen anomaly in which right valve of embryonic sinus venosus persists and extends from right atrial wall toward interatrial septum, with fibrous membrane dividing the right atrium into 2 compartments.[2] If this structure persists as filaments and fibrils, it is called Chiari network.[3]

Cor triatriatum was first defined by Church in 1898, and echocardiographic diagnosis was made in 1984 by Ostman-Smith et al.[3] Distribution of clinical findings differs based on obstructive characteristics (if any) of the fibromuscular membrane of the right atrium. Patients may present with right heart failure or supraventricular arrhythmias, or they may be completely asymptomatic.[4] Fesslova et al. evaluated previously published cases and reported on case diagnosed by fetal echocardiogram during intrauterine life. This newborn underwent surgery during postnatal period due to desaturation of 75% to 85%.[5] Mackman et al. published series of 3 cases with neonatal cyanosis. Most patients have either slightly decreased peripheral oxygen saturation or they are completely asymptomatic.[6] Our patient had no known complaint; echocardiogram performed with initial diagnosis of ARF carditis revealed fibromuscular membrane in the right atrium near the VCS-right atrium junction. Our patient was hemodynamically stable, and no sign of obstruction was detected.

CTD can accompany structural abnormalities of the right heart, such as tricuspid valve anomalies, pulmonary artery stenosis or atresia, atrial septal defect, and Ebstein anomaly.[7,8] In the present patient, no concomitant cardiac abnormality other than ARF carditis-associated first-degree mitral failure and trace residual aortic regurgitation was found.

In follow-up of cases of cor triatriatum, and especially in cases of CTS, surgical treatment is frequently required. Decision to operate in case of CTD is based on clinical findings. In asymptomatic CTD cases without heart failure, arrhythmia, or concomitant cardiac anomaly, clinical observation is recommended as sufficient. Our patient was asymptomatic, and clinical monitoring has been maintained.

Generally, transthoracic echocardiogram suffices to make diagnosis of CTD. However, as reported in the literature, some cases have been diagnosed during catheterization or angiographic examination.[8] The disease has no specific physical examination or laboratory finding that supersedes meticulous application of echocardiography. This report of 12-year-old asymptomatic patient who received diagnosis of CTD has been presented to emphasize importance of echocardiography so as not to overlook such a rarely seen cardiac anomaly.

Authorship contributions

Concept: A.O., H.A.G., İ.İ.Ç.; Design: H.A.G., A.O.; Data collection &/or processing: A.E.K., E.A.; Analysis and/or interpretation: A.O., H.A.G.; Literature search: A.O., A.E.K., E.A.; Writing: A.O., H.A.G.; Critical review: İ.İ.Ç., H.A.G.

Conflict of interest

None declared.

REFERENCES

1. Zainudin AR, Tiong KG, Mokhtar SA. Cor triatriatum dexter: A rare cause of childhood cyanosis. Ann Pediatr Cardiol 2012;5:92–4.
2. Bejiqi R, Retkoceri R, Bejiqi H, Zeka N, Kelmendi M. Echocardiographic diagnosis of divided right atrium-cor triatriatum dextrum.

Med Arh 2010;64:303–4.
3. Çiftçi H, Kılıç Z, Angı E, Ünalır A, Timuralp B. İki Boyutlu Ekokardiyografi ile Saptanan Anormal Bir Sağ Atrial Oluşum: "Chiari Network". Turkiye Klinikleri J Cardiol 1988;1:156–7.
4. Kouchoucos NT, Blackstone EH, Doty DB, Stanley FL, Karp RB, editors. Cor triatriatum. In: Kirklin/Barratt-Boyes cardiac surgery. 3rd ed. Philadelphia: Elsevier Science; 2003. p. 781–9.
5. Fesslova V, Saracino A, Nuri H, Pomé G. Cor triatriatum dexter: unusual features in utero and after birth. Interact Cardiovasc Thorac Surg 2012;14:330–2. [CrossRef]
6. Mackman CA, Liedel JL, Woods RK, Samyn MM. A case series of patients with cor triatriatum dexter: unique cause of neonatal cyanosis. Pediatr Cardiol 2015;36:240–3. [CrossRef]
7. Embrey RP. Cor triatriatum, pulmonary vein stenosis, atresia of the common pulmonary vein. In: Mavroudis C, Backer CL editors. Pediatric Cardiac Surgery. 2nd ed. St Louis: Mosby-Year Book; 1994. p. 503–4.
8. Gürsu HA, Varan B, Özkan M, Erdoğan İ, Oktay A, Aşlamacı S. Cor triatriatum: a single institution's experience. Türk Göğüs Kalp Damar Cerrahisi Dergisi 2015;23:9–13. [CrossRef]

Nadir Bir Olgu: Kor Triatriatum Dekster

Kor triatriatum dekster (KTD) sinüs venosusdaki embriyonal sağ kapağın sebat etmesi sonucu oluşan oldukça nadir görülen bir doğumsal kalp defektidir. Kor triatriatum deksterin klinik bulguları tıkanmanın derecesi ile ilişkilidir. Bazı semptomsuz olgular farklı nedenlerle ekokardiyografi çekilirken tanı alabilirler. Bu yazıda, akut romatizmal ateş (ARA) ön tanısı ile çekilen ekokardiyogramda KTD saptanan 12 yaşındaki kız hasta sunuldu. Bu sunumun amacı, KTD tanısının zor olduğunu ve tanı için ekokardiyogram ile sağ atriumun dikkatli bir şekilde incelenmesi gerektiğini vurgulamaktı.

Anahtar Sözcükler: Ekokardiyografi; kor triatriatum; romatizmal ateş; üç boyutlu.