CASE REPORT

Case Report: Fortuitous discovery of cor triatriatum sinister in a young adult [version 1; peer review: 1 not approved]

Achour Asma ¹, Mohamed Kacem Ben Slima¹, Chamtoury Ikram ²,
Aida Ammar¹, Mabrouk Abdelali ¹, Jamel Saad¹, Badii Hmida¹, Ahmed Zrig¹,
Mejdi Ben Massoud ², Walid Mnari¹, Mezri Maatouk¹

¹Radiology Department, Fattouma Bourguiba University Hospital, Faculty of Medicine, Monastir University, Monastir, 5000, Tunisia

²Cardiology Department, Fattouma Bourguiba University Hospital, Faculty of Medicine, Monastir University, Monastir, 5000, Tunisia

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Abstract

Cor triatriatum is an extremely rare cardiac congenital malformation characterized by subdivision of the left atrium into two chambers. This division is due to the presence of an abnormal fibromuscular membrane interrupted by one or more openings that communicate the two chambers. The size and number of those orifices is variable, affecting the age of onset of symptoms and their severity. Our case describes the fortuitous discovery of this pathology in a young adult in her forties, following the finding of atrial fibrillation during a preoperative assessment of a strangulated hernia. The cardiac ultrasound allowed in our case to invoke the diagnosis, confirmed thereafter with an angioscanner. Surgical resection was not required in our patient who was put on medical treatment involving a beta-blocker to reduce atrial fibrillation and an anticoagulant treatment to decrease the thromboembolic risk, with a good clinical evolution. It seems therefore interesting to consider this uncommon pathology which may be latent for a long time, manifesting after some physiological or pathological situations.

Keywords

Cor triatriatum, defect, heart, cardiac CT-scan, sinister, dexter, case report

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1

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[view](#)

1. **Nagarajan Muthialu** , Great Ormond
Street Hospital, London, UK

Any reports and responses or comments on the article can be found at the end of the article.

Corresponding author: Achour Asma (doc.asma.achour@hotmail.com)

Author roles: **Asma A:** Conceptualization, Formal Analysis, Writing – Original Draft Preparation; **Ben Slima MK:** Conceptualization; **Ikram C:** Investigation; **Ammar A:** Visualization, Writing – Original Draft Preparation; **Abdelali M:** Visualization; **Saad J:** Visualization; **Hmida B:** Visualization; **Zrig A:** Supervision, Visualization; **Ben Massoud M:** Methodology; **Mnari W:** Validation; **Maatouk M:** Methodology, Supervision, Validation

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Case presentation

A 44-year-old G5P5 white female with no medical or surgical history and with no cardiovascular risk factors, who was a teacher and a mother of five, consulted the emergency room for occlusive syndrome and acute abdominal pain. She had no particular pathological history particularly with no cardiovascular disease risk factors. The diagnosis of a strangulated umbilical hernia was made, requiring emergency surgery. Physical examination revealed an irregular tachycardia at 150 beats per minute, a blood pressure of 120/70 mmHg, with a normal respiratory and neurological examination. An electrocardiogram as part of the preoperative assessment showed the presence of persistent atrial fibrillation previously unknown.

A cardiac echography was made showing a dilated left atrium containing a septum dividing it into two chambers. Continuous wave Doppler allowed to locate the orifice within the accessory membrane without significant pressure gradient. The left ventricle was not dilated with a preserved ejection fraction. A cardiac CT-scan confirmed the diagnosis and allowed the visualization of an accessory chamber measuring 11.5 cm in diameter (Figure 1), receiving all pulmonary veins, and communicating with the left atrium via a large single opening in the membrane, three centimeters in size (Figure 2). No thrombus was detected, and no signs of pulmonary hypertension (Figure 3).

Figure 1. Cardiac CT-scan: 4-chamber view (a) and 2-chamber view (b).

Figure 2. Single large opening at the upper part of the membrane.

Figure 3. Normal pulmonary trunk and pulmonary arteries diameters.

Open surgical resection was not performed in view of the absence of obstructive symptoms and the absence of pulmonary congestion symptoms.

Atrial fibrillation was treated with beta-blocker by injection of 5 to 10 mg (1 mg/min) of Atenolol (Tenormine®) followed by an oral dose of 50 mg. The patient was subsequently put on a vitamin K antagonist with an initial dose of warfarin of 4 mg orally once a day followed by a maintenance dose of 7 mg orally once a day for life. The patient was discharged after two weeks with a regular sinus rhythm and a good general state. At three months follow-up, the patient kept a regular sinus rhythm with no worsening of the intra-atrial pressure gradient on ultrasound control.

Our case seems unique because of the latency of the symptomatology despite the occurrence of several factors of decompensation, in particular the five pregnancies, explained by the width of the communication within the intra-atrial septum estimated at 3 cm.

In fact, this pathology generally becomes symptomatic in adulthood following several physiological or pathological factors of decompensation¹ or following the appearance of fibrosis or calcification of the accessory membrane orifice.² It is therefore important to supervise asymptomatic non-operated patients, as in our case, by regular clinical and ultrasound examination to prevent possible decompensation.

Discussion

Cor triatriatum is a rare congenital heart defect, representing 0.1 to 0.4% of all cardiac malformations,¹ which can be isolated, or in up to 80% of cases^{2,3} associated with many cardiac malformations, most commonly atrial septal defect, followed by a patent foramen ovale and left superior vena cava. Less frequently, it is associated with anomalous pulmonary venous return, mitral valve regurgitation, isolated pulmonary artery stenosis, and tetralogy of Fallot.^{2,3}

Two forms have been described: Cor triatriatum Sinister (CTS) in which the fibromuscular septum sits in the left atrium, and Cor triatriatum Dexter (CTD), rarer than CTS, interesting the right atrium.²

Cor triatriatum sinister, which corresponds to the case reported, was first described by Church in 1868.⁴ The embryonic cause remains not clearly established, but the most widely approved is malin corporation theory which consists of failure of the common pulmonary vein to fuse with the left atrium creating a complete, incomplete, or fenestrated septum.⁵

Classification of CTS includes 3 types: A, B and, C. The case described here matches the type A.²

Patients commonly develop symptoms early in life,¹ during childhood or adolescence, or later in adulthood, or being completely asymptomatic throughout the whole life, depending on the number and size of orifices in the membrane.¹ The most severe presentations can cause neonatal respiratory distress with an increased mortality risk.¹ However, adults with the disease are usually asymptomatic.²

Some physiological and pathological situations including all causes of increased cardiac flow and venous return may precipitate symptoms. Pregnancy is a common example associated with tachycardia and increased cardiac flow may cause cardiac decompensation in asymptomatic patients.⁶ In this case, the patient was in her forties at diagnosis and already had five pregnancies without any incidents reported, which is explained by the large communication between the two chambers minimizing hemodynamic implications.

A chest x-ray as initial investigation may find pulmonary congestion (Kerley B-lines, acute pulmonary edema, pleural effusion), cardiomegaly with left atrial enlargement and prominent pulmonary vessels.⁷

ECG is most likely normal or may show unspecific features such as right axis deviation, S1Q3 pattern, or atrial arrhythmia.^{1,2}

A cardiac ultrasound done for rhythm disorders is usually the circumstance of discovery. It allows visualizing the membrane, number, and size of the openings, to estimate the transmembrane pressure gradient, flow velocity and to detect complications (such as thrombosis) and associated malformations.²

Recently, the diagnosis performance of this technique has been improved by the contribution of 3D reconstruction of transesophageal US scan which shows exact membrane morphology.⁸

Classically, diagnosis is made by cardiac angiography. Nowadays diagnosis is confirmed by a CT or MR scan.² Magnetic Resonance Imaging (MRI) has the advantage of a finer study by showing more details. CT scan offers better spatial resolution and the possibility of multiplanar reconstructions.

Treatment depends on the clinical presentation. The conservative approach is preferred in asymptomatic presentations.⁹

Thus, asymptomatic patients without pressure gradient do not require medical treatment, but in case of signs of pulmonary overload a medical treatment is proposed in addition to the surgical treatment.¹ Some successful catheter ablation assays were also reported.²

Medical treatment targets hemodynamic stabilization, cardiac rhythm control and anticoagulation for patients with atrial fibrillation.⁷

Surgery is the definitive treatment. It consists of atriotomy, excision of interatrial membrane, and correction of associated anomalies.¹ The result of surgical repair seems to be excellent in most the cases.²

Our case report demonstrates the latency of this pathology depending on the size of the accessory membrane orifice. Indeed, this pathology may remain asymptomatic for a long time with a risk of being revealed at an adult age following certain situations, by a cardiac decompensation or a thromboembolic complication. The width of the orifice within the accessory membrane explains in our case the delay of the diagnosis in spite of five pregnancies which might have been responsible for a cardiac decompensation.

The rarity of this pathology and the lack of specificity of the clinical signs could orient us wrongly towards more frequent pathologies such as mitral stenosis, but the diagnosis is quickly rectified by the cardiac echography or secondarily by an angioscanner or an MRI. The prognosis after surgical treatment seem to be excellent according to a review of the literature,³ hence the interest of not delaying the positive diagnosis, avoiding the risk of sudden death to the patients.

Data availability

All data underlying the results are available as part of the article and no additional source data are required.

Consent

Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

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Nagarajan Muthialu

Department of Cardiothoracic Surgery, Great Ormond Street Hospital, London, UK

Many thanks for asking me to review this case report.

While it is interesting to note the incidental finding and diagnosis of cor triatriatum in an adult, it is not a rare congenital defect warranting publication as a single case report.

Additionally (and more importantly), I have concerns on the diagnosis itself. The images do not show enough evidence of this being haemodynamically significant - the membrane is not shown fully on the slices of echocardiographic pictures. Even if they were to be shown, I wonder if conservative management is appropriate. The patient in question is in mid 40s age group. The risk of atrial arrhythmias, and consequent thromboembolic phenomenon will only worsen with time, and in the presence of this membrane, it is further accentuated, while we may not know the actual risk for this.

With these, I would think this is not appropriate for indexing in its current form.

Is the background of the case's history and progression described in sufficient detail?

No

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Yes

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

No

Is the case presented with sufficient detail to be useful for other practitioners?

No

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Paediatric cardiothoracic surgery

I confirm that I have read this submission and believe that I have an appropriate level of expertise to state that I do not consider it to be of an acceptable scientific standard, for reasons outlined above.

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