CASE REPORT

Cor triatriatum sinistrum: presentation of syncope and atrial tachycardia

Malcolm Avari,¹ Sunil Nair,² Zofia Kozłowska,² Samer Nashef³¹James Paget University Hospitals NHS Foundation Trust, Great Yarmouth, UK²Department of Cardiology, James Paget University Hospitals NHS Foundation Trust, Great Yarmouth, UK³Papworth Hospital NHS Foundation Trust, Cambridge, UK

Correspondence to

Dr Malcolm Avari, malcolmavari@hotmail.com

Accepted 11 January 2017

SUMMARY

We present a rather unusual cause for syncope associated with atrial tachycardia. A man aged 39 years presented with an episode of syncope and narrow complex tachycardia. Further investigations, including transoesophageal echocardiography, identified cor triatriatum sinistrum (CTS), a rare congenital abnormality characterised by the atrium being divided by a fibrous membrane. Although it is rare, there has been an increase in diagnosis due to developments in diagnostic imaging techniques. Symptoms are related to the size of fenestrations within the fibrous membrane. Presenting symptoms can mimic those seen in mitral stenosis. It is a condition that can occur in isolation, but it can also be associated with other cardiac abnormalities such as an atrial septal defect (ASD) (as in this case). Surgery is the definitive treatment (this man had surgical repair of CTS and closure of ASD) and should be considered at any age if there are any associated symptoms or complications.

BACKGROUND

This case highlights the features of a rare congenital cardiac anomaly which can be found in adults and should be considered as a differential diagnosis when patients present with supraventricular tachycardias, heart failure or features similar to mitral stenosis. Although it is rare, there has been an increase in diagnosis due to developments in diagnostic imaging techniques.

CASE PRESENTATION

A fit and well man aged 39 years with a background of asthma (on inhalers) presented with an episode of syncope. Earlier on the day of admission, while standing up from kneeling on the floor, the patient went pale and light-headed but did not lose consciousness. Later that day, when parking his car, he suddenly blacked out and bumped into two parked cars. Prior to this episode, he experienced no chest pain or shortness of breath but had noticed palpitations. He had no relevant social or family history. On physical examination, a splitting of second heart sound was heard. The patient was advised to stop driving at work as a plant operator until fully investigated for cardiac issues.

INVESTIGATIONS

While in the emergency department, his ECG showed supraventricular tachycardia-atrial tachycardia with 1:1 conduction at 220 bpm (figure 1). He was given 300 mg of amiodarone which caused reversion back to sinus rhythm. His subsequent ECGs showed T wave inversion in leads V1–V3 and

right axis deviation. A transthoracic echocardiogram (TTE) performed showed right ventricular (RV) hypertrophy and abnormal flow towards the RV apex. Transoesophageal echocardiography (TOE) later performed confirmed RV hypertrophy with normal size and good function. On multiple views, a clear linear structure was seen to divide the left atrium just on the atrial side of the mitral valve (fibrous membrane)—cor triatriatum sinistrum (CTS) (figure 2). There were two high velocity flows across the membrane (fenestrations) connecting the two left atrial chambers (video 1). The main pulmonary artery beyond the pulmonary valve appeared dilated. The left ventricular size and function appeared normal.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses that should be considered are atrial tumours, pulmonary arterial hypertension, mitral stenosis and supraventricular tachycardias.

TREATMENT

In this case, as the patient was symptomatic (atrial arrhythmia) with features on echocardiogram suggesting compromise, the patient was initially started on verapamil 240 mg and referred for eventual cardiac surgery.

OUTCOME AND FOLLOW-UP

At the previous follow-up, postsurgical repair of CTS and closure of atrial septal defect (ASD) performed in 2015, the patient had experienced a couple of episodes of palpitations lasting few seconds only, with no further episodes of syncope. The patient is continuing under yearly medical follow-up.

DISCUSSION

Cor triatriatum (CT) was first described by Church in 1868 in a woman aged 38 years.^{1 2} It is a rare congenital condition where the incidence is 0.1–0.4% with a ratio of men:women affected being 1.5:1 and no genetic predisposition.^{3 4} This condition usually involves the left atrium (CTS) compared with the right atrium (CT dexter).⁵ The left atrium is divided into two chambers by a fibromuscular membrane resulting in ‘true left atrium’ where the anterior-inferior chamber associates with the left ventricle via mitral valve and the posterior-superior chamber where the four pulmonary veins join.^{4 5}

Three main theories have been stipulated regarding CTS: malseptation (membrane dividing the left atrium is an abnormal growth of septum primum), malincorporation (incomplete fusion of the

CrossMark

To cite: Avari M, Nair S, Kozłowska Z, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-218395

Figure 1 ECG showing narrow complex tachycardia–atrial tachycardia with 1:1 conduction block (rate 220 bpm).

Figure 2 Transoesophageal echocardiogram showing CTS. (A) (i) Fibromuscular septum; (ii) mitral valve. DC, distal chamber; PC, proximal chamber; LV, left ventricle. (B) Doppler flow showing mitral regurgitation.

embryonic common pulmonary vein into the left atrium) and entrapment (left horn of the sinus venosus entraps the common pulmonary vein and prevents its incorporation into the left atrium).^{3 6} In 1949, Loeffler classed CT into three subgroups depending on the size and number of openings within the fibrous membrane: group 1 (no opening), group 2 (one or more small openings) and group 3 (wide opening). Those patients falling in group 3 are more likely to develop no or few symptoms of the disease where the condition is diagnosed incidentally and are likely to survive beyond infancy into late adulthood.^{3 6 7} In this case, our patient falls into group 3 category. Survival is related to the size of fenestration in the fibrous membrane and ~50% of patients die in infancy.⁸ Generally, when the fenestration diameter is >1 cm, patients are symptom-free as there is no intra-atrial pressure gradient.^{5 9} It is not fully understood why patients with large fenestrations within the fibrous membrane become symptomatic as adults. Electronic and mechanical remodelling of the myocardial tissue occur and it is thought that calcification or fibrosis of the membrane's opening can contribute to narrowing, flow obstruction and atrial fibrillation, which can increase the risk of thromboembolic events.^{3 7 10}

As well as CTS occurring in isolation (classic), there have been other congenital cardiac associations linked with CTS (atypical). Humpl *et al* showed that the majority of patients

Video 1 Transoesophageal echocardiogram video showing fibromuscular septum with regurgitation with Doppler imaging.

(77%) presented with associated cardiac lesions when data from 1954 to 2005 of 82 patients with CTS were analysed. They also found that the most common associations with CTS were ASD (53%), patent ductus arteriosus (18%) or partial or total anomalous pulmonary venous drainage (27%).^{1 3 10 11} There has also been a case report of the rare association of CTS with bicuspid aortic valve.¹²

ECG may be normal in presentations of CTS but can often demonstrate atrial arrhythmias (atrial tachycardia and atrial fibrillation)

Learning points

- It is important to raise awareness of a rare congenital condition such as cor triatriatum sinistrum (CTS) due to its potential treatment and consider CTS as a differential diagnosis when patients present with supraventricular tachycardias, heart failure or symptoms that mimic mitral stenosis.^{8 10}
- It is a condition that can occur in isolation or with other congenital cardiac associations.^{1 3}
- Imaging modalities such as echocardiogram, cardiac MRI and computed tomography can be used for identification.^{3 4}
- Intervention, usually by means of definitive cardiac surgery, is considered if there are associated symptoms or complications.

and RV hypertrophy.^{5 9} There are several imaging modalities that can be used to diagnose CTS such as TTE, TOE, computed tomography, cardiac MRI and cardiac catheterisation.^{3 4 11}

For those patients with CTS as an incidental finding and who are asymptomatic, no treatment is usually required. However, if at any age symptoms (such as dyspnoea or orthopnoea, like in mitral stenosis) are present as a result of the obstructive function of the intra-atrial membrane, surgical treatment is the definitive course of therapy.^{4 11} Surgical treatment consists of resection of the membrane and repair of associated cardiac defect through median sternotomy after establishing cardiopulmonary bypass, as in this case.² Survival is >90% at 5 years postoperation with improvement of symptoms.¹¹

Contributors MA performed literature search, author and guarantor. SuN involved in identifying and managing the case, provided idea for article and clinic letters, author and guarantor. SaN contributed to management of the case and provided operation notes. ZK provided reports of investigations performed.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Humpl T, Reineker K, Manlhoit C, *et al.* Cor triatriatum sinistrum in childhood. A single institution's experience. *Can J Cardiol* 2010;26:371–6.
- 2 Slight RD, Nzewi OC, Sivaprakasam R, *et al.* Cor triatriatum sinister presenting in the adult as mitral stenosis. *Heart* 2003;89:e26.
- 3 Chen Q, Cuhathakurta S, Vadalapali G, *et al.* Cor triatriatum in adults: three new cases and a brief review. *Tex Heart Inst J* 1999;26:206–10.
- 4 Kokotsakis J, Anagnostakou V, Almpanis G, *et al.* Cor triatriatum presenting as heart failure with reduced ejection fraction: a case report. *J Cardiothorac Surg* 2011;6:83.
- 5 Nassar PN, Hamdan RH. Cor triatriatum sinistrum: classification and imaging modalities. *Eur J Cardiovasc Med* 2011;1:84–7.
- 6 Loeffler E. Unusual malformation of the left atrium: pulmonary sinus. *Arch Pathol* 1949;48:371–6.
- 7 Strickland PT, Pernetz MA, Jokhadar M, *et al.* Cor triatriatum sinister: a patient, a review, and some unique findings. *Echocardiography* 2014;31:790–4.
- 8 Jorgensen CR, Ferlic RM, Varco RL, *et al.* Cor triatriatum. Review of the surgical aspects with a follow-up report on the first patient successfully treated with surgery. *Circulation* 1967;36:101–7.
- 9 Modi KA, Annamali S, Ernest K, *et al.* Diagnosis and surgical correction of cor triatriatum in an adult: combined use of transoesophageal and contrast echocardiography, and a review of literature. *Echocardiography* 2006;23:506–9.
- 10 Eichholz JL, Hodroge SS, Crook JJ II, *et al.* Cor triatriatum sinister in a 43-year-old man with syncope. *Tex Heart Inst J* 2013;40:602–5.
- 11 Alphonso N, Norgaard MA, Newcomb A, *et al.* Cor triatriatum: presentation, diagnosis and long-term surgical results. *Ann Thorac Surg* 2005;80:1666–71.
- 12 Schusterova I, Czerny M, Jurko A Jr, *et al.* A very rare case of late diagnosis of cor triatriatum sinistrum. *J Geriatr Cardiol* 2015;12:185–6.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit

<http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow