CASE REPORT

Idiopathic aneurysmal giant right atrial enlargement with thrombus formation

Rashi Khare,¹ Sharad Chandra,² Vikas Agarwal,¹ Sudhanshu Dwivedi²

¹King George's Medical University, Lucknow, Uttar Pradesh, India

²Department of Cardiology, King George's Medical University, Lucknow, Uttar Pradesh, India

Correspondence to

Dr Rashi Khare,
rashimedicine@yahoo.com

Accepted 6 December 2014

SUMMARY

A 12-year-old boy presented to our outpatient department with progressive dyspnoea and palpitations of 6-month duration. Cardiovascular system examination revealed a short ejection systolic murmur at the left lateral heart border. Chest X-ray showed gross cardiomegaly. Two-dimensional ECHO revealed massive aneurysmal right atrial (RA) enlargement, low-pressure tricuspid regurgitation and dysplastic tricuspid valve with compression of the left atrium (LA) and left ventricle (LV). Cardiac CT showed grossly enlarged RA compressing the LA, LV, right pulmonary artery and right upper lobe pulmonary vein. Initially, the patient refused surgery. On follow-up, a thrombus was seen in the giant RA and the patient agreed to and underwent successful surgery. Absence of pericardium was specifically excluded on cardiac MRI and on surgery. Resected atrial tissue showed wall thinning and focal fibrosis establishing idiopathic dilation of the RA. The patient is on regular follow-up.

BACKGROUND

Aneurysmal massive enlargement of right atrium (RA) without any underlying pathology is an uncommon entity and has been rarely reported. Our case report highlights the clinical importance of this condition and describes the widely available treatment.

CASE PRESENTATION

A 12-year-old boy presented to our outpatient department with progressive dyspnoea and palpitation of 6-month duration. Cardiovascular system examination revealed a short ejection systolic murmur at the left lateral heart border. Chest X-ray showed gross cardiomegaly (figure 1). Two-dimensional ECHO revealed a massive 11.3×9.7 cm aneurysmal RA enlargement (figure 2), low pressure tricuspid regurgitation (figure 3) and dysplastic tricuspid valve (figures 4 and 5). Ebstein's anomaly was specifically excluded by measuring the apical displacement of the tricuspid valve and the mitral valve (figure 6). The massively enlarged RA was compressing the left atrium (LA) and left ventricle (LV) with preserved biventricular function. Cardiac CT showed grossly enlarged RA compressing the LA, LV, right pulmonary artery and right upper lobe pulmonary vein (figures 7 and 8). Initially, the patient refused surgery and was managed conservatively. On follow-up, thrombus was seen in the giant RA (figure 9) and the patient agreed to and underwent successful tricuspid valve repair with RA reduction surgery. Absence of pericardium was specifically excluded on cardiac MRI and on surgery. Resected atrial tissue showed wall thinning and focal fibrosis

Figure 1 Chest X-ray showing huge cardiomegaly, with right atrial enlargement.

establishing idiopathic dilation of the RA. The patient is on regular follow-up and is asymptomatic.

DIFFERENTIAL DIAGNOSIS

Ebstein's anomaly.

OUTCOME AND FOLLOW-UP

The patient is on regular follow-up and is asymptomatic.

DISCUSSION

Idiopathic RA enlargement is a very rare entity. It has been defined as an increase in the right atrial long axis indexed to body surface area (men

Figure 2 Two-dimensional ECHO subcostal four-chamber view showing a giant 11.3×9.7 cm right atrium.

To cite: Khare R, Chandra S, Agarwal V, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-207559

Figure 3 Two-dimensional ECHO four-chamber view with continuous wave doppler (CW) showing low-pressure tricuspid regurgitation.

Figure 4 Two-dimensional ECHO showing the dysplastic tricuspid valve.

Figure 5 Three-dimensional ECHO showing dysplastic tricuspid valve and giant right atrium (RA).

Figure 6 Two-dimensional ECHO four-chamber view showing 7 mm difference between apical displacement of mitral and tricuspid valve, hence excluding Ebstein's anomaly.

Figure 7 Cardiac CT showing giant right atrium compressing left atrium and left ventricle.

Figure 8 Cardiac CT showing giant right atrium compressing right pulmonary artery.

$>2.6 \text{ cm}^2$, women $>2.8 \text{ cm}^2$) in the absence of other cardiac abnormalities.¹ It can be congenital or acquired. Most patients are asymptomatic with some presenting with dyspnoea, chest pain, fatigue and palpitation.² Atrial fibrillation, atrial

Figure 9 Follow-up three-dimensional ECHO showing thrombus in giant right atrium (RA).

flutter and preexcitation have been reported as some causes of palpitation. Because of sluggish blood flow in the giant RA, these patients are prone to thrombus formation, which can lead to thromboembolic complications. Prolonged atrial fibrillation

can also lead to similar complications.³ Exclusion of Ebstein's anomaly and absence of pericardium are very important for making the diagnosis of idiopathic RA enlargement.^{4 5} RA reduction surgery with tricuspid valve (TV) repair is the preferred treatment strategy.

Learning points

- ▶ Aneurysmal idiopathic right atrial (RA) enlargement can be a cause of massive cardiomegaly.
- ▶ Ebstein's anomaly and absence of pericardium must be excluded to make the diagnosis of idiopathic RA enlargement.
- ▶ These patients are prone to thrombus formation.
- ▶ Accurate diagnosis is important as the condition can be easily treated by RA reduction surgery.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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