Coronary arteriovenous fistula with coexisting atrial septal defect

Saraschandra Vallabhajosyula,¹ Pranathi Rao Sundaragiri,¹ Pallavi Bellamkonda,² Mark Jeffrey Holmberg²

¹Department of Internal Medicine, Alegant-Creighton University Medical Center, Creighton University School of Medicine, Omaha, NE, USA
²Division of Cardiovascular Medicine, Department of Internal Medicine, Alegant-Creighton University Medical Center, Creighton University School of Medicine, Omaha, NE, USA

Correspondence to

Dr Saraschandra Vallabhajosyula, SaraschandraVallabhajosyula1@creighton.edu

Accepted 24 March 2014

DESCRIPTION

A 36-year-old Hispanic woman was referred for an asymptomatic cardiac murmur and auscultation revealed wide and fixed second heart sound. Transthoracic echocardiogram showed severely dilated right atrium and ventricle. Transesophageal echocardiogram (TEE) confirmed left-to-right shunt through secundum atrial septal defect (ASD) (1.5 cm × 2.0 cm × 2.5 cm), with Qp:Qs 2:1 on Doppler studies (figure 1). In view of large ASD size and inadequate superior rim of tissue, the patient was planned for surgical repair. Routine coronary angiogram showed a significant coronary arteriovenous fistula (CAVF) from the sinoatrial branch of the right coronary artery to the common pulmonary artery (PA; figure 2). The patient underwent autologous pericardial patch repair of ASD

with CAVF ligation, with no post-operative complications.

CAVF is a congenital coronary anomaly noted in 0.002% of the general population and 0.08–0.30% of angiographic series, hypothesised to be due to persistent sinusoidal connections between the coronary arteries and primitive heart tubules.^{1–3} In 20–45% cases, other associated congenital cardiac anomalies are evident.⁴ Drainage is primarily into the right heart system (90%) with 15–17% PA drainage.^{2–4} Complications include angina (coronary steal), heart failure, myocardial infarction and death.^{1–4} Concurrent ASD is reported to have a significantly higher Qp/Qs ratio.² Despite coronary angiogram being the gold standard of diagnosis, alternate modalities such as TEE, CT angiogram and MRI are increasingly being advocated.^{3–4}

Figure 1 Three-dimensional view on transesophageal echocardiogram demonstrating atrial septal defect.

To cite: Vallabhajosyula S, Sundaragiri PR, Bellamkonda P, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-203500

Figure 2 Coronary angiogram (left anterior oblique view) demonstrating coronary arteriovenous fistula from right coronary artery.

Learning points

- Recognising rare coronary anomalies such as coronary arteriovenous fistula (CAVF) with emphasis on diagnostic modalities.
- Highlighting the rare association of CAVF with other congenital cardiac conditions and the challenges faced in management.

Contributors SV was involved in case workup and management, literature review, drafting and finalising the manuscript. PRS contributed by drafting and finalising the manuscript. PB was involved in case workup and management and finalising the manuscript. MJH contributed by critically reviewing, analysing and finalising the manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Dimitrakakis G, Von Oppell U, Luckraz H, *et al*. Surgical repair of triple coronary-pulmonary artery fistulae with associated atrial septal defect and aortic valve regurgitation. *Interact Cardiovasc Thorac Surg* 2008;7:933–4.
- 2 Sunder KR, Balakrishnan KG, Tharakan JA, *et al*. Coronary artery fistula in children and adults: a review of 25 cases with long-term observations. *Int J Cardiol* 1997;58:47–53.
- 3 Jonjev ZS, Milosavljevic A, Redzek A, *et al*. Successful repair of coronary artery-pulmonary artery fistula with coronary artery disease and atrial septal defect in an adult. *J Thorac Cardiovasc Surg* 2005;130:890–1.
- 4 Singhal P, Liang M, Devlin G, *et al*. Congenital left main coronary artery to main pulmonary artery fistula with bicuspid aortic valve: a case report and review of literature. *J Card Surg* 2010;25:295–9.

Copyright 2014 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