A rare case of pulmonary valve infective endocarditis in a patient with ventricular septal defect

Raja Ezman Faridz Raja Shariff, Sazzli Shahlan Kasim and Hafisyatul Aiza Zainal Abidin

Abstract

Right-sided infective endocarditis (IE) is rare and often affects the tricuspid valve. We report a unique case of pulmonary valve IE in a patient with a predisposing congenital heart defect – a ventricular septal defect (VSD). A 23-year old man with a VSD was admitted following 3 months’ history of fever and malaise. An initial transthoracic echocardiogram (TTE) failed to reveal any visible vegetations or mass. However, blood cultures revealed persistent methicillin-sensitive *Staphylococcus aureus* (MSSA). A transoesophageal echocardiogram (TOE) showed multiple hyperechoic structures in the entirety of the anterior cusp of the pulmonary valve, suggestive of vegetations. In view of his young age and subacute presentation, a trial of prolonged antibiotics was opted for. The patient was commenced on intravenous cloxacillin for 6 weeks, which was successful. Common risk factors for right-sided IE include intravenous drug abuse, central venous catheterization and alcoholism. Less common risk factors include left-to-right shunts, including VSD. Proposed mechanisms include turbulent jet flow causing damage to the valve and vegetation formation. Although response to antibiotics and prognosis in right-sided IE tend to be better than in left-sided IE, surgical intervention may still be indicated, and unfortunately, evidence remains scarce on the appropriate patient selection for surgical intervention. Isolated pulmonary valve IE due to predisposing VSD remains a rare entity. It is important to consider this diagnosis in prolonged pyrexia of unknown origin in individuals with known congenital heart defects.

Keywords

Infective endocarditis, ventricular septal defect, case report

Introduction

Right-sided infective endocarditis (IE) is uncommon (5–10% of total IE) and often affects the tricuspid valve. Only 1.5–2% of hospital admissions involving IE affect the pulmonary valves. We report a unique case of pulmonary valve IE in a patient with a predisposing congenital heart defect – a ventricular septal defect (VSD).

Case report

A 23-year-old man, known to have a VSD since the age of 3 years, was transferred to our centre from a neighbouring hospital for further investigations following intermittent episodes of fever and malaise over a 3-month period. The patient denied any recent travelling, hospitalization or abuse of illegal intravenous substances. The patient suffered from untreated, severe acne and admitted to touching his skin using unwashed hands at times. Aside from low-grade pyrexia (37.8 Celsius), vital signs on arrival were stable, including an oxygen saturation of 98% on room air. Clinical examination revealed both a grade 2 pansystolic and a grade 3 early dias- tolic murmur.

Initial transthoracic echocardiography (TTE) by the referring hospital revealed dilated right atrium and ventricles; a 0.8 cm perimembranous, restrictive VSD, with a visible shunt on colour Doppler and VSD jet velocity of 4 m/s; and a pulmonary-to-systemic flow (Qp:Qs) ratio of 1.7. Their calculated pulmonary artery systolic pressure was 45 mmHg, and pulmonary artery mean pressure was 30 mmHg. However, no visible vegetations or mass was demonstrated. Due to raised inflammatory markers (white cell count of 28 × 10^12/l and C-reactive protein of 230 mg/ml) and detection of methicillin-sensitive *Staphylococcus aureus* (MSSA), a transoesophageal echocardiogram (TOE) showed multiple hyperechoic structures in the entirety of the anterior cusp of the pulmonary valve, suggestive of vegetations. In view of his young age and subacute presentation, a trial of prolonged antibiotics was opted for. The patient was commenced on intravenous cloxacillin for 6 weeks, which was successful. Common risk factors for right-sided IE include intravenous drug abuse, central venous catheterization and alcoholism. Less common risk factors include left-to-right shunts, including VSD. Proposed mechanisms include turbulent jet flow causing damage to the valve and vegetation formation. Although response to antibiotics and prognosis in right-sided IE tend to be better than in left-sided IE, surgical intervention may still be indicated, and unfortunately, evidence remains scarce on the appropriate patient selection for surgical intervention. Isolated pulmonary valve IE due to predisposing VSD remains a rare entity. It is important to consider this diagnosis in prolonged pyrexia of unknown origin in individuals with known congenital heart defects.
Staphylococcus aureus (MSSA) on blood cultures, the patient was started on intravenous cloxacillin for 2 weeks. However, due to persistently raised inflammatory markers and persistent MSSA in blood cultures on three sets of sampling each 72 hours apart, a decision was made to transfer the patient to our centre for transoesophageal echocardiography (TOE). TOE revealed the earlier-mentioned VSD and additionally demonstrated an abnormal pulmonary valve morphology, specifically of the anterior cusp, with evidence of anterior cusp prolapse. There appeared to be multiple hyperechoic structures in the entirety of the anterior cusp, the largest measuring 0.5 cm × 0.6 cm in size (Figure 1). TOE was also able to demonstrate two different aliasing colour flow Dopplers, one throughout the systolic phase, representing the VSD left-to-right shunt, and one during the early diastolic phase, representing likely severe pulmonary regurgitation flow (Figure 2). Colour flow Doppler did not reveal any valvular cusp perforations, although no three-dimensional imaging of the valves was available to ascertain perforations or the exact morphology of the mass for accurate measurements.

Early surgical consult was sought due to worries about potential complications. However, in view of his young age and subacute presentation, a trial of prolonged antibiotics was opted for. The patient was on intravenous cloxacillin for a total of 6 weeks. There was subsequent improvement in clinical state and blood investigations. Repeat TTE and TOE demonstrated clearance of any masses, although the anterior cusps of the pulmonary valve showed evidence of persistent prolapse with severe pulmonary regurgitation. The patient, however, remained asymptomatic. He was subsequently referred to the cardiothoracic team, and underwent both VSD closure and pulmonary valve replacement in another institution.

Discussion

Right-sided IE is relatively rare in comparison to left-sided IE. Reasons for this include the lower transvalvular pressure gradients and venous oxygen concentration in right chambers, as well as lower rates of congenital defects and malformations seen in the right side of the heart. However, there may be an under-estimation of the true prevalence of concurrent right-sided IE and adult congenital heart disease, as very little exists in the current literature.

Common risk factors for right-sided IE include intravenous drug abuse (30%), central venous catheterization (14%) and alcoholism (11%). Less common risk factors include left-to-right shunts, including patent ductus arteriosus and VSD, although VSD is more commonly associated with tricuspid valve IE. Proposed mechanisms of right-sided IE with co-existing shunts include turbulent jet flow, causing shear stress, and circumferential stretch, which damages the endothelial cells of the valves. The endothelial disruption causes fibrin deposition and consequent vegetation formation.

Often, the vegetations tend to localize on the ventricular side of a pulmonary valve IE (in the path of the regurgitation) and on the atrial side of a tricuspid valve IE. In our case, due to the infundibular location of VSD and pre-existing pulmonary valve prolapse, the vegetations were...
seen in the side nearer to the right ventricular outflow tract (RVOT).

Response to antibiotics and prognosis in right-sided IE tend to be better than in its left-sided counterpart. However, surgical intervention may still be indicated, specifically when right heart failure, persistent infection despite antibiotics, large-sized vegetations and recurrent pulmonary embolism are present.1–6 Unfortunately, evidence remains scarce on the appropriate patient selection for surgical intervention in isolated pulmonary valve involvement. In our case, the patient was referred to the cardiothoracic team only after IE clearance, as there were clear indications for VSD closure, due to the Qp:Qs ratio, and possible benefit from simultaneous repair of the pulmonary valve prolapse.

**Conclusion**

Isolated pulmonary valve IE due to predisposing VSD remains a rare entity. It is important to consider this diagnosis in prolonged pyrexia of unknown origin in individuals with known congenital heart defects. Our case highlights the importance, when assessing VSD patients with persistent pyrexia for possible IE, of looking for vegetations not just surrounding the VSD site but further downstream at the RVOT, where the jet hits the infundibulum, as well as the pulmonary valve.

**Acknowledgements**

The authors would like to acknowledge Universiti Teknologi MARA (UiTM) for supporting the submission of the following article.

**Authors’ contributions**

- REFRS: Data collection and analysis, drafting of manuscript
- SSK: Drafting of manuscript, revision of manuscript
- HAZA: Drafting of manuscript, revision of manuscript

**Availability of data and materials**

The data that support the findings of this study are available from UiTM Sungai Buloh, but restrictions apply to the availability of these data, which were used under licence for the current study and so are not publicly available. Data are, however, available from the authors upon reasonable request and with permission of UiTM Sungai Buloh.

**Ethical approval**

Ethical approval to report this case was obtained from the Universiti Teknologi MARA (UiTM) Ethics Committee (Ethics Committee Registration Number 07/2020). The manuscript does not report on any animal data or tissue.

**Informed consent**

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

**Conflict of interest**

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

**Funding**

The authors received no financial support for the research, authorship, and/or publication of this article.

**ORCID iD**

Raja Ezman Faridz Raja Shariff [https://orcid.org/0000-0002-5167-5863]

**References**

1. Park HE, Cho GY, Kim HK, et al. Pulmonary valve endocarditis with septic pulmonary thromboembolism in a patient with ventricular septal defect. J Cardiovasc Ultrasound 2009; 17: 138–140.
2. Kumar B, Singh A, Akram M, et al. Nature’s balancing act: infective endocarditis of pulmonary valve with ventricular septal defect in fifth decade: a rare and unusual presentation. J Cardiol Cases 2017; 17: 77–79.
3. Nishida K, Fukuyama O and Nakamura DS. Pulmonary valve endocarditis caused by right ventricular outflow obstruction in association with sinus of Valsalva aneurysm in a patient with ventricular septal defect. J Cardiothorac Surg 2008; 3: 46.
4. Fazlinezhad A, Fallah A and Esfahanizadeh J. Pulmonic valve endocarditis with pulmonary artery endarteritis in a young man with congenital ventral septal defect. ARYA Atheroscler 2010; 6: 42–44.
5. Turhan O, Saba R, Belgi A, et al. A case of right-side infective endocarditis with ventricular septal defect. Infez Med 2005; 13: 39–41.
6. Sattwikka PD, Hartopo AB, Anggrahini DW, et al. Right-sided infective endocarditis in patients with uncorrected ventricular septal defect and patent ductus arteriosus: two case reports. Clin Case Rep 2018; 6: 2168–2173.