CASE REPORT

A case of residual inferior sinus venosus defect after ineffective surgical closure

Sayuri Uga,¹ Takayuki Hidaka,¹ Taiichi Takasaki,² Yasuki Kihara¹

¹Department of Cardiovascular Medicine, Hiroshima University Graduate School of Biomedical and Health Sciences, Hiroshima, Japan

²Department of Surgery, Hiroshima University Graduate School of Biomedical and Health Sciences, Hiroshima, Japan

Correspondence to

Dr Sayuri Uga,
gaugaugao@hiroshima-u.ac.jp

Accepted 9 September 2014

SUMMARY

A 38-year-old woman presented with cyanosis and heart failure 34 years after patch closure of an atrial septal defect and partial anomalous pulmonary venous connection. CT and cardiac catheterisation showed a residual defect that caused right-to-left shunting. The patch almost blocked the inferior vena cava from the right atrium, resulting in uncommon drainage of the inferior vena cava into the left atrium. Other anomalies included the coronary-to-pulmonary artery fistula and duplicate inferior vena cava with dilated azygos venous system. A second surgery was performed, and we confirmed an inferior sinus venosus defect, which is rare and can be misdiagnosed. The ineffective patch closure had caused a haemodynamic status that rarely occurs. We describe the diagnostic process and emphasise the importance of correctly understanding the entity.

BACKGROUND

An inferior sinus venosus defect (SVD) is a rare type of atrial septal defect (ASD) that could be mistaken for a secundum ASD. Misdiagnosis can lead to poorer outcomes. The present case underwent additional surgical repair 34 years after the first surgery because of a residual defect. Accurate pre-operative diagnosis is important to determine an appropriate surgical strategy.

CASE PRESENTATION

A 38-year-old woman presented with extraordinary hypoxia, cyanosis and heart failure following a miscarriage. At the age of 4 years, she had undergone patch closure of an ASD and partial anomalous pulmonary venous connection (PAPVC) in another hospital. Although she had inexplicable hypoxia after the operation, she led a normal life except for intolerance to intense exercise.

Physical examination revealed the following: blood pressure 98/50 mm Hg, normal lung auscultation and a 2/6 diastolic murmur along the left sternal border. Jugular venous distension and ankle oedema were not visible, while cyanosis and a clubbed finger were observed.

Peripheral oxygen saturation was 90% at rest and decreased to 60% during the 6 min walk test, at which point she experienced mild dyspnoea. The oxygen saturation remained at 93% under sufficient oxygen supplementation.

INVESTIGATIONS

Laboratory findings revealed arterial hypoxaemia and an alveolar-arterial oxygen difference of 59.8 mm Hg (table 1). ECG showed sinus

bradycardia and a negative T wave in the right precordial leads. Chest radiography revealed an enlarged heart shadow with increased pulmonary vasculature. Echocardiography showed dilation of the left atrium (LA) and left ventricle with preserved systolic function. Right-to-left shunting was not detected by contrast echocardiography using agitated saline injected into the arm.

DIFFERENTIAL DIAGNOSIS

The arterial blood gas analysis suggested the presence of ventilation-perfusion mismatch due to a pulmonary embolism or a right-to-left shunting as the mechanism for hypoxaemia. Echocardiography showed no change in the right ventricular morphology associated with acute pulmonary embolism or Eisenmenger syndrome. We were unable to detect right-to-left shunting in the contrast echocardiography and lung perfusion scintigraphy.

However, CT disclosed a residual defect causing right-to-left shunting (figure 1), and we established the diagnosis of inferior SVD. The inferior vena cava (IVC) was almost sealed off from the right atrium (RA) by the patch, resulting in diversion of the blood flow from the IVC into the LA. In addition, CT showed a coronary-to-pulmonary artery fistula, right upper pulmonary vein obstruction and duplicate IVC (figure 2) with dilated azygos venous system. The left-sided IVC joined the retro-aortic left renal vein and continued to the haemiazzygos-azygos veins, which ended in the superior vena cava (SVC).

Cardiac catheterisation revealed a step up in the blood oxygen saturation at the pulmonary artery (PA) and a step down at the left ventricle. The coronary-to-pulmonary artery fistulas from both

Table 1 Preoperative and postoperative blood gas analysis relating to surgery for a residual inferior sinus venosus defect

	Preoperative	Postoperative
pH	7.401	7.423
PCO ₂ (mm Hg)	29.5	40.4
PO ₂ (mm Hg)	53.3	105
HCO ₃ ⁻ (mmol/L)	17.9	25.9
BE (mmol/L)	-5.2	1.8
Lactate (mmol/L)	0.9	1.0
O ₂ saturation (%)	84.7	98.4
A-aDO ₂	59.8	

A-aDO₂, alveolar-arterial oxygen difference; BE, base excess; PCO₂, partial pressure of carbon dioxide; PO₂, partial pressure of oxygen.

CrossMark

To cite: Uga S, Hidaka T, Takasaki T, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-205377

Figure 1 Preoperative CT revealing right-to-left shunting due to a residual inferior sinus venosus defect. (A) The previous patch (arrow) separates the RA from the IVC. (B) The defect continues from the IVC to the LA without an inferior septal margin. (C) The sagittal section showing the overridden IVC that is connected almost entirely to the LA. RA, right atrium; LA, left atrium; LV, left ventricle; IVC, inferior vena cava.

coronary arteries to main PA were well grown and caused coronary artery steal.

TREATMENT

Owing to these findings, open-heart surgery was performed. The previous patch was removed and the posterior border of the atrial septum was incised. Then, the atrial septum was reconstructed using a 0.4 mm thick prosthetic patch (Gore-Tex patch, W. L. Gore & Associates, Inc, Flagstaff, Arizona, USA). The coronary-to-pulmonary artery fistula was closed by suturing to control the inflows into the main PA.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful, and the follow-up CT 11 days after the operation showed that the new patch that was sutured to the atrial septum completely covered the defect (figure 3). The patient was discharged 2 weeks after the operation.

DISCUSSION

SVDs, which were first identified by Ross in 1956,¹ are an uncommon class of ASDs and are often associated with PAPVC of the right pulmonary veins. They are characterised by malposition of the insertion of the SVC or IVC, which overrides the atrial septum,² and results in extraseptal communication between the right and left atria. Inferior SVDs are particularly rare, leading to difficulties with preoperative diagnoses. Misdiagnosis has been associated with worse outcomes^{3 4}; therefore, an accurate diagnosis is important. Transoesophageal echocardiography is useful for detecting inferior SVDs and PAPVC.⁵

It is particularly important to distinguish inferior SVDs from secundum ASDs with inferior extension in order to determine

the appropriate therapeutic strategy. With inferior SVDs, the defect originates in the mouth of the IVC and continues directly into the inferoposterior border of the LA, leaving no residual atrial septal tissue at the inferior margin.⁶ However, with secundum ASDs with inferior extension, a rim of septal tissue always exists between the ASD and mouth of the IVC. They are rarely associated with PAPVC.

Some cases with iatrogenic diversion of IVC to the LA after surgical closure of ASD have been reported.⁷ This rare complication can happen when the Eustachian valve is mistaken for the inferior rim of ASD in the repair of low-lying ASDs and inferior SVDs. Although we obtained no evidence that the presence of the Eustachian valve was related to the condition in the present case, we concluded that the inappropriate patch closure led to rare haemodynamics that would be observed with direct drainage of the IVC into the LA. Therefore, we were unable to detect right-to-left shunting by the usual method of injecting a contrast agent into the arm during an echocardiography and lung perfusion scintigraphy. We would certainly detect the shunting by injection into the femoral vein. In the present case, the CT and cardiac catheterisation were useful in diagnosing the inferior SVD and other anomalies that accounted for the manifestation.

Different approaches for the treatment of inferior SVDs have been successfully reported, including a suture closure with⁸ and without⁹ a patch, and reimplantation of the anomalous pulmonary vein.¹⁰ We need to look for the entry point of the IVC cannula into the IVC and suturing the patch beneath the cannula not to mistake the Eustachian valve for the inferior margin of ASD. In the patch closure, it is necessary to look for the entry point of the IVC cannula into the IVC and suture the patch beneath the cannula not to mistake the Eustachian valve for the inferior margin of ASD.

Figure 2 Preoperative cardiac catheterisation and CT showing additional findings in this case with a residual inferior sinus venosus defect. (A) Coronary angiogram in left anterior oblique 60 view showing coronary-to-pulmonary artery fistulas (arrows) originate from the RCA and drain to the main PA. Fistulas from RCA and LAD were well grown. On CT scan, (B) right upper pulmonary vein obstruction (arrow) and (C) duplicate inferior vena cava (arrow) were observed. RCA, right coronary artery; PA, pulmonary artery; LAD, left anterior descending coronary artery.

Figure 3 Postoperative CT showing that (A) a Gore-Tex patch (arrows) covered the defect, and (B and C) the patch (arrows) was sutured continuously from the posterior wall of the IVC to the atrial septum. LA, left atrium; IVC, inferior vena cava; RA, right atrium.

We think the coronary-to-pulmonary artery fistula in this case was an acquired anomaly that occurred after the first surgery because no fistula was noted in the records from the first surgery. We believe that the dilated azygos venous system and coronary-to-pulmonary artery fistula might compensate for the right-to-left shunting from the IVC to the LA.

The direct drainage of the IVC to the LA can cause life-threatening manifestations such as cyanosis as well as the paradoxical embolism. That is why we need to be thoroughly aware of this rare complication after surgery of ASD. We describe this rare case to emphasise the need for accurate diagnosis to enable appropriate treatment.

Learning points

- ▶ An inferior sinus venous defect (SVD) is a rare type of atrial septal defect that can be misdiagnosed.
- ▶ We experienced a rare case of inferior SVD that had uncommon drainage of the inferior vena cava into the left atrium caused by a previous surgery.
- ▶ CT and cardiac catheterisation were useful in detecting the residual right-to-left shunting and other anomalies.
- ▶ It is important to understand the entity for accurate diagnosis and effective treatment.

Acknowledgements The authors thank Professor Taijiro Sueda (Department of Surgery, Hiroshima University Graduate School of Biomedical and Health Sciences) for his encouragement; Dr Yoshihiro Dohi (Department of Cardiovascular Medicine) for his assistance with the investigation; and all colleagues for contribution to this work.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Shaner RF. The high defect in the atrial septum. *Can Med Assoc J* 1958;78:688–90.
- 2 al Zaghaf AM, Li J, Anderson RH, *et al*. Anatomical criteria for the diagnosis of sinus venosus defects. *Heart* 1997;78:298–304.
- 3 Banka P, Bacha E, Powell AJ, *et al*. Outcomes of inferior sinus venosus defect repair. *J Thorac Cardiovasc Surg* 2011;142:517–22.
- 4 Yilmaz K, Ewert P, Hetzer R, *et al*. Surgical repair after ineffective device closure of an inferior sinus venosus defect. *Interact Cardiovasc Thorac Surg* 2012;14:485–7.
- 5 Kronzon I, Tunick PA, Freedberg RS, *et al*. Transesophageal echocardiography is superior to transthoracic echocardiography in the diagnosis of sinus venosus atrial septal defect. *J Am Coll Cardiol* 1991;17:537–42.
- 6 Plymale J, Kolinski K, Frommelt P, *et al*. Inferior sinus venosus defects: anatomic features and echocardiographic correlates. *Pediatr Cardiol* 2013;34:322–6.
- 7 Jain SA, Pinto R, Dalvi B. Iatrogenic diversion of IVC to left atrium after surgical closure of ASD. *Ann Pediatr Cardiol* 2012;5:72–4.
- 8 Crystal MA, Al Najashi K, Williams WG, *et al*. Inferior sinus venosus defect: echocardiographic diagnosis and surgical approach. *J Thorac Cardiovasc Surg* 2009;137:1349–55.
- 9 Sturm JT, Ankeney JL. Surgical repair of inferior sinus venosus atrial septal defect. *J Thorac Cardiovasc Surg* 1979;78:570–2.
- 10 Perna AM, Alajmo F. Partial anomalous pulmonary venous return of an uncommon type. *J Cardiovasc Surg* 1984;25:563–5.

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