Unroofed coronary sinus and persistent left superior vena cava

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Received 13 February 2006; received in revised form 26 May 2006; accepted 2 June 2006
Available online 11 July 2006

Abstract This report describes a case of unroofed coronary sinus and persistent left superior vena cava discovered during an echocardiographic investigation for dilated pulmonary artery. An unroofed coronary sinus is a rare interatrial shunt that is commonly associated with a persistent left superior vena cava. The latter is a usual cause of a dilated coronary sinus. The detection of a dilated coronary sinus should therefore prompt the search for abnormal coronary sinus drainage and other cardiac abnormalities. The treatment of unroofed coronary sinus and persistent left superior vena cava is undertaken only after assessing the pre- and post-treatment haemodynamics of all co-existing abnormalities.

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Introduction

The complex of an unroofed coronary sinus (UCS) and a persistent left superior vena cava (PLSVC) is a rare congenital heart disease first described by Raghib et al. in 1965.1 A normal coronary sinus drains the cardiac veins into the right atrium. A UCS, in addition to draining the cardiac veins, also communicates abnormally with the left atrium. This abnormal communication is thought to be due to impaired development of the partition between the left atrium and the coronary sinus—an alternative explanation is subsequent dissolution of this partition.2 A PLSVC, abnormally draining the left internal jugular and subclavian veins into the coronary sinus, is due to impaired degeneration of the embryonic left counterpart of the normal right superior vena cava.3 A UCS or a PLSVC may be further associated with other cardiac abnormalities.

Case study

A 65-year-old woman presented with haemoptysis attributed to chest infection. She did not have breathlessness or cyanosis and the haemoptysis resolved. Her chest X-ray was suggestive of a dilated right pulmonary artery subsequently confirmed with a thoracic computed tomography. A bronchoscopy ruled out bronchial malignancy. Her electrocardiograph showed right bundle branch block with normal axis.

She was referred for an echocardiographic evaluation of her right heart. A transthoracic echocardiography showed a left-to-right shunt from the left atrium into a dilated UCS. The atria, right ventricle and main pulmonary artery were also dilated (Figs. 1–3). The right ventricle showed volume overload with diastolic flattening of the interventricular septum and the pulmonary artery systolic pressure was mildly raised. The biventricular systolic function was good.

Normal saline agitated with patient’s blood was injected into patient’s left antecubital vein. Bubble contrast appeared in the coronary sinus before appearing in the right heart thus confirming...
a PLSVC (Fig. 4). Some bubble contrast also appeared in the left atrium consistent with some right-to-left shunt but the shunt is mainly left-to-right. Not all the pulmonary veins were visualised and associated thoracic venous abnormalities such as anomalous pulmonary venous drainage could not be ruled out.

She therefore had a transoesophageal echocardiography that showed partial anomalous pulmonary venous drainage (PAPVD) with the right pulmonary veins emptying into the superior vena cava. The left pulmonary veins were drained normally. A repeat saline bubble contrast study confirmed the PLSVC (Fig. 5). No atrial or ventricular septal defect was seen. The echocardiographer detected colour flow from the left atrium into the coronary sinus but could not detect the UCS per se.

The patient proceeded to having a right cardiac catheter that crossed aberrantly into the left atrium and left ventricle signifying an interatrial shunt not detected on the transoesophageal echocardiography. A repeat analysis of the transoesophageal echocardiography revealed crossing of some bubbles from the coronary sinus into the left heart. The UCS was therefore detected on the transthoracic echocardiography and right cardiac catheterisation but not fully on the transoesophageal echocardiography. The right cardiac catheterisation also revealed a pulmonary artery systolic pressure at upper limit of normal, shunt ratio of 2.2:1, normal pulmonary vascular resistance and step-up in the oxygen saturation at the level of high right atrium.
She was referred to our regional centre for grown-up congenital heart disease and was treated conservatively with annual echocardiography. The patient remains well two years after her index haemoptysis.

Discussion

UCS and PLSVC may cause no symptoms or may cause right ventricular failure,4 paradoxical cerebral embolism and cerebral abscess,5 or cyanosis that may vary with neck position.6 In this case report, the haemoptysis is attributed to chest infection but may also be related to the increased pulmonary blood flow from the shunt.

A PLSVC is typically suspected when aberrant catheterisation of the pulmonary artery is demonstrated on chest X-ray after catheterisation via the left internal jugular or subclavian veins.3 If a UCS is also present, catheterisation of the left heart and aorta is possible7 and can therefore cause spurious ‘pulmonary hypertension’, aortic vascular injuries and embolism.

UCS and PLSVC may be further associated with other cardiac abnormalities such as atrioventricular septal defect, atrial appendage anomalies and coronary sinus ostial atresia.8 This case is associated with a PAPVD. UCS, PLSVC and associated cardiac abnormalities may be investigated with echocardiography,9 cardiac magnetic resonance imaging10 cardiac computed tomography or cardiac catheterisation.

Treatment of UCS and PLSVC, if needed, is surgical correction of its components and associated abnormalities. For example, the PLSVC may be occluded percutaneously5,6 or re-routed surgically to the right heart circulation such as the left pulmonary artery.11 Treating the PLSVC alone may not be sufficient — the UCS may need to be patched and other associated abnormalities treated accordingly.

These procedures should be performed only after anticipating their haemodynamic effects on the coronary sinus drainage and associated abnormalities. This is to avoid disturbed venous return or residual shunt8 leading to risky re-operations. For example, the normal innominate vein drains the left internal jugular and subclavian veins into the normal right superior vena cava. In its absence, a PLSVC becomes the sole drainage for these left neck and arm veins and should be re-routed and not occluded11 to prevent congestion of the head and upper limb.

Conclusion

Dilated coronary sinus is a prompt to look for further cardiac abnormalities such as intracardiac shunts or thoracic venous abnormalities. The complex of UCS and PLSVC is one such abnormality and its treatment requires careful assessment of not only the UCS and PLSVC but also other concomitant cardiac abnormalities to prevent post-treatment haemodynamic complications.

References

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Echocardiography remains the standard for the diagnostic evaluation of left ventricular tumors: A case report with anatomical correlation

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Received 27 March 2006; received in revised form 2 June 2006; accepted 15 June 2006
Available online 28 July 2006

KEYWORDS
Cardiac tumors; Papillary fibroelastoma; Echocardiography; Contrast echocardiography; Cardiac magnetic resonance; Stroke

Abstract Primary cardiac tumors are rare. Although the majority are benign, they may cause significant morbidity and mortality. Two-dimensional transthoracic echocardiography (2D-TTE) is the primary imaging modality for the diagnosis of cardiac tumors. New and more complex non-invasive imaging modalities, such as cardiac magnetic resonance (CMR), do not always provide an added value. This is illustrated in the presented case report of a papillary fibroelastoma (PFE).

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Case report

A 78-year-old woman with known arterial hypertension was admitted for cerebellar stroke with acute gait disturbances and left limb ataxia. Her ECG showed sinus rhythm.

The two-dimensional transthoracic echocardiography (2D-TTE) revealed an echo-dense, homogeneous, round, pedunculated and highly mobile (diameter approximately 14 mm) structure attached to the apex of the left ventricle (Fig. 1). A finding consistent with a cardiac tumor.