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Brief Report

Absence of the coronary sinus with coronary venous drainage into the main pulmonary artery

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Abstract We describe the case of a 9-year-old girl demonstrating isolated absence of the coronary sinus with abnormal coronary venous drainage into the main pulmonary artery. Coronary angiography showed normal coronary arterial trees and contrast medium from both coronary arteries drained into the main pulmonary artery via an abnormal cardiac vein on the anterior wall of the right ventricle.

Keywords: Absence of coronary sinus; coronary sinus; cardiac vein

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Atresia of the coronary sinus ostium and absence of the coronary sinus are relatively rare cardiac malformations and may be associated with other congenital heart diseases including single ventricle, hypoplastic left heart syndrome, and atrial septal defect.[1] According to a population-based database, 2.3% of patients with univentricular heart are associated with atresia of the coronary sinus ostium.[2] Most cases of atresia of the coronary sinus ostium have persistent left superior caval vein with retrograde flow from the persistent left superior caval vein into the innominate vein or with unroofed coronary sinus.[1] In a few instances, atresia of the coronary sinus ostium or absence of the coronary sinus is accompanied by Thebesian veins draining directly into the cardiac chambers or caval veins.[3] To the best of our knowledge, there are no previously reported cases of isolated absence of the coronary sinus with coronary venous drainage into the main pulmonary artery.

Case report

A 9-year-old girl had been diagnosed at 2 months of age as having mild pulmonary valve stenosis. Electrocardiogram did not show any abnormal findings during follow-up. She was referred to Saitama Children's Medical Center because of removal from her family. On physical examination, 3/6 ejection systolic murmur and early diastolic murmur were detected at the left upper sternal border. Chest radiography did not show any signs of either cardiomegaly or dilatation of the pulmonary artery. Two-dimensional echocardiography showed mild dilatation of the main pulmonary artery and colour Doppler echocardiography demonstrated a mosaic pattern in the main pulmonary artery; however, the pulmonary valve was not thickened and its opening was not restricted.

Therefore, she was admitted for cardiac catheterisation. Right heart catheterisation showed pulmonary artery pressure of 32/18 mmHg and right ventricular pressure of 39 mmHg at systole. There was no left-to-right shunt detected on oxygen saturation. Normal systemic and pulmonary venous drainage were demonstrated on conventional angiographic injection. The levoangiogram phase of right ventriculography demonstrated opacification of the main pulmonary artery. Selective coronary artery injections showed normal coronary arterial trees; however, during the venous phase, contrast from both coronary arteries drained into the abnormal cardiac vein on the anterior wall of the right ventricle (Figs 1 and 2). There were small arteriovenous anastomoses between the right coronary artery and the cardiac vein. The cardiac vein began at the apex of the heart and coursed upward

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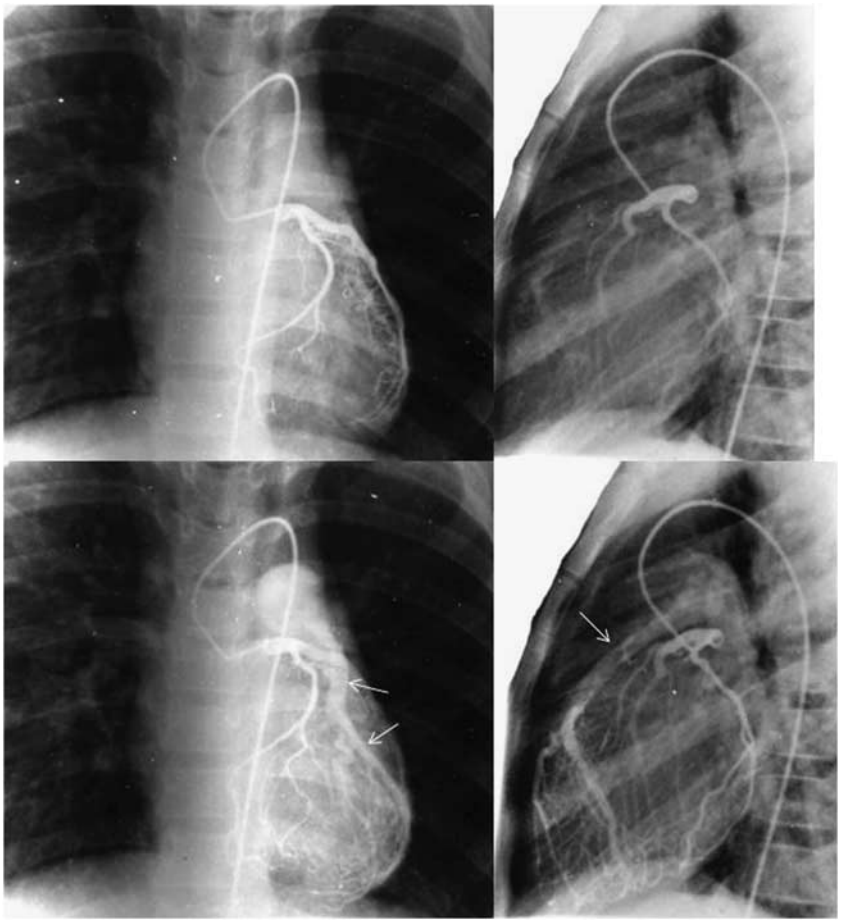

Figure 1.
Angiogram of the left coronary artery (frontal and lateral view). Upper: normal coronary arterial trees, lower: venous phase. Arrow indicates abnormal cardiac vein.

along the left anterior descending artery until midpoint of the anterior groove. It subsequently turned off to the right and emptied into the main pulmonary trunk. The normal coronary sinus was not visualised during the venous phase after selective coronary artery injections.

She was followed up over the course of 6 years and complained of chest pain during exercise at 15 years of age. Treadmill exercise testing showed myocardial ischaemia. Therefore, we advised her to participate in low-intensity competitive sports only.

Discussion

Malformations of the coronary sinus are rare congenital diseases, including enlargement, hypoplasia, absence, atresia of the coronary sinus ostium, and partial or complete unroofing.[4,5]

Atresia of the coronary sinus ostium is always combined with persistent left superior caval vein draining the coronary sinus retrograde to the innominate vein; a gross connection from the coronary sinus to the left atrium (unroofed coronary sinus); drainage of the coronary sinus directly into the cardiac chambers via Thebesian veins; or persistence of a levoatrio-cardinal vein connecting the coronary sinus and the left atrium. There may be several diseases associated with any of the above.[6]

Absence of the coronary sinus is extremely rare, and it may be difficult to discriminate between absence and atresia of the coronary sinus in patients with other congenital heart defects.[5] Absence of the coronary sinus is also usually associated with other abnormalities, most commonly persistent left superior caval vein draining into the left atrium with atrial septal defect (infero-posterior type).[7] In such cases, the cardiac veins (Thebesian veins) drain individually into their corresponding atria. Foale et al[8] reported a case of complete absence of the coronary sinus occurring as an isolated abnormality, in which the cardiac veins drained separately into the left and right atrium. Bergman et al[9] reported a heart specimen in which the coronary sinus was absent and the great cardiac veins drained into the

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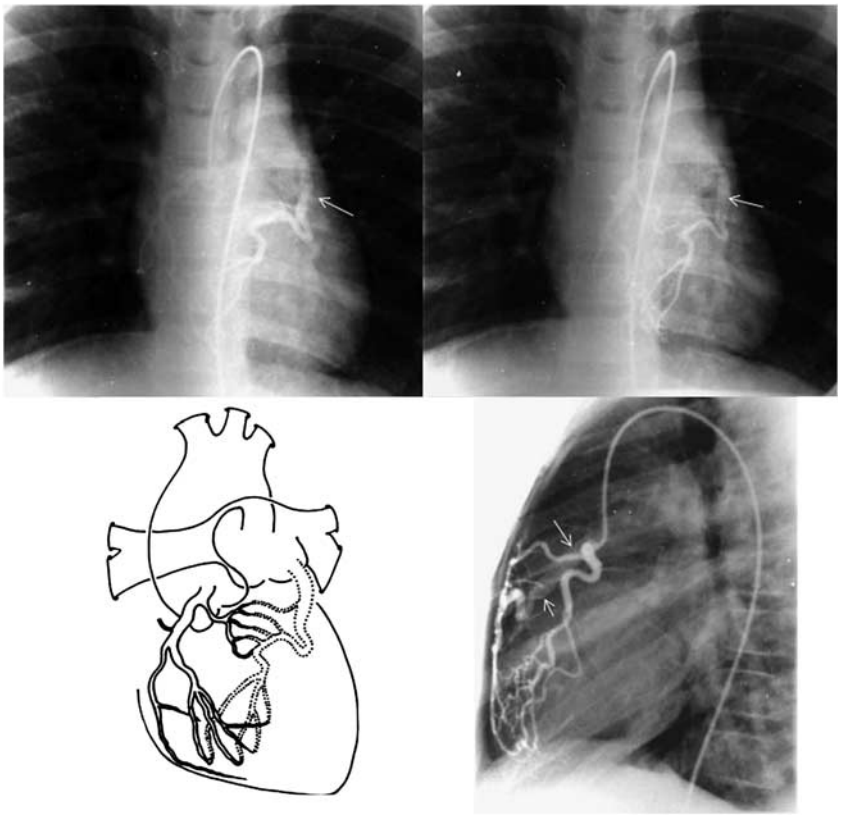

Figure 2.
Angiogram of the right coronary artery (frontal and lateral view). The coronary venous blood from the right coronary artery drained into the abnormal cardiac vein. There were small arteriovenous anastomoses. Arrow indicates abnormal cardiac vein.

right superior caval vein and right atrium. Rao et al[10] reported a 42-year-old man with isolated absence of the coronary sinus and cardiac venous drainage into the left ventricle through multiple coronary-cameral fistulous connections.

To the best of our knowledge, there are no previously reported cases of isolated absence of the coronary sinus with coronary venous drainage into the main pulmonary artery. In our case, coronary venous blood drained into the abnormal cardiac vein on the anterior wall of the right ventricle. The cardiac vein originated at the apex of the heart and coursed upward along the left anterior descending artery to the mid-point of the anterior groove. Therefore, the abnormal cardiac vein might be the anterior interventricular vein with aberrant course. Our case is similar to Von Lüdighausen's case. Von Lüdinghausen described a heart specimen showing an aberrant course of the anterior interventricular vein in which the vein originated in the distal anterior interventricular sulcus. After leaving the sulcus and continuing towards the right, the vein passed over the arterial conus and the root of the pulmonary trunk.[6]

The reason why the arteriovenous fistula developed only between the right coronary artery and the vein is not understood well. This difference might be relevant to the fact that collateral channels develop significantly in the right coronary artery in patients with obstructive coronary artery lesions in the chronic phase of Kawasaki disease.

Atresia or absence of the coronary sinus is not considered clinically important because it is physiologically corrected as the coronary venous blood enters the pulmonary circulation. Recognition of this abnormality has generally been considered of vital importance only to cardiac surgeons during the repair of associated cardiac lesions.[1] However, a 42-year-old man reported by Rao et al[10] complained of dyspnoea on exertion and showed ST segment depression on treadmill test. Our case was also associated with chest pain during exercise and showed myocardial ischaemia on treadmill exercise testing. Myocardial perfusion might be adversely affected by exercise because the coronary venous blood returned to the higher pressure site than the right atrium.

References

1. Janssen DR, Graham TP, Doyle TP, Moore JD. Occlusion of the coronary sinus ostium: a danger for cavopulmonary anastomosis. Catheter Cardiovasc Interv 2005; 64: 108–111.

https://doi.org/10.1017/S1047951112001709 Published online by Cambridge University Press

2. Salminen JT, Hakala T, Pihkala J, et al. Coronary sinus orifice atresia with left superior vena cava in patients with univentricular heart. Ann Thorac Surg 2006; 81: e16–e17.
3. Lucus RV Jr, Krabill KA. Abnormal systemic venous connections. In: Emmanouilides GC, Reimenschneider TA, Allen HD (eds.). Heart Disease in Infants, Children, and Adolescents, 5th edn. Williams & Wilkins, Baltimore, 1995, pp 874–902.
4. Mantini E, Grondin CM, Lillehei CW, Edwards JE. Congenital anomalies involving the coronary sinus. Circulation 1966; 33: 317–327.
5. Adatia I, Gittenberger-De Groot AC. Unroofed coronary sinus and coronary sinus orifice atresia. Implications for management of complex congenital heart disease. J Am Coll Cardiol 1995; 25: 948–953.
6. von Lűdinghausen M. The venous drainage of the human myocardium. Adv Anat Embryol Cell Biol 2003; 168: 1–104.
7. Raghib G, Ruttenberg HD, Anderson RC, et al. Termination of left superior vena cava in left atrium, atrial septal defect, and absence of coronary sinus. Circulation 1965; 31: 906–918.
8. Foale RA, Baron DW, Rickards AF. Isolated congenital absence of coronary sinus. Br Heart J 1979; 42: 355–358.
9. Bergman RA, Thompson SA, Saadeh FA. Absence of the coronary sinus. Anat Anz 1988; 166: 9–12.
10. Rao CV, Raghu K, Sharada K, Jaishankar S. Absent coronary venous sinus: a rare anomaly. Indian Heart J 2001; 53: 352–353.

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