Case Report

Isolated persistent left-sided superior vena cava, giant coronary sinus, atrial tachycardia and heart failure in a child

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A R T I C L E  I N F O

Article history:
Received 4 October 2012
Accepted 10 August 2013
Available online 26 August 2013

Keywords:
Atrial tachycardia
Congestive heart failure
Coronary sinus
Persistent LSVC

A B S T R A C T

Persistence of a left-sided superior vena cava (PLSVC) with absent right superior vena cava (isolated PLSVC) is a very rare venous malformation and commonly associated with congenital heart disease or alterations of the cardiac situs. We describe an unusual case of a young boy presenting with persistent atrial tachycardia and congestive heart failure. He was detected to have unexplained grossly dilated right atrium, right ventricle with systolic dysfunction and a giant coronary sinus (CS). The dilated CS closely mimicked a pseudo cor-triatriatum on echocardiography. Contrast echocardiography from both arms revealed opacification of the CS before the right atrium. Bilateral upper limb venography confirmed the presence of absent right SVC and isolated persistent left SVC draining into the giant coronary sinus.

1. Introduction

Persistence of a left-sided superior vena cava (PLSVC) is the most common variant of systemic venous drainage. In most patients with PLSVC, a right sided superior vena cava is present which drains normally into the right atrium. A PLSVC with absent right SVC (Isolated PLSVC) is an extremely rare entity and commonly associated with congenital heart disease or alterations of the cardiac situs. We describe an unusual case of a young boy who presented with persistent atrial tachycardia, aneurysmally dilated right atrium, right ventricular systolic dysfunction and a hugely dilated coronary sinus (CS). The dilated CS closely mimicked a cor-triatriatum on echocardiography. On injection of agitated saline from both arms, opacification of the CS occurred before the right atrium. Bilateral upper limb venography was performed which demonstrated the presence of absent right SVC, a bridging vein and isolated PLSVC draining into the hugely dilated CS. A high index of suspicion, bubble contrast echocardiography and/or bilateral antecubital venous contrast venography is very helpful in discovery of this uncommon venous anomaly.

2. Case summary

The patient was JS, a 14-year-old young male who presented with palpitations, gradually progressive shortness of breath, abdominal distension and swelling feet of 3 months duration. At presentation, he was tachypneic with a pulse rate of...
214/min which was regular and low volume. The jugular venous pressure was elevated with prominent a and v waves; gross ascites, tender pulsatile hepatomegaly, bilateral pedal edema and an audible pansystolic murmur along lower sternal border suggestive of tricuspid regurgitation (TR), was noted.

A 12 lead ECG (Fig. 1) showed a narrow QRS tachycardia suggestive of atrial tachycardia at a rate of 214/min with 1:1 conduction. Chest radiograph showed gross cardiomegaly with minimal right sided pleural effusion. Trans-thoracic echocardiography revealed situs solitus, aneurysmally dilated right atrium (RA), dilated right ventricle (RV) with moderate RV systolic dysfunction. Moderate tricuspid regurgitation (TR) was noted with mildly elevated RV systolic pressure (25 mm Hg). The left sided chambers including the mitral and aortic valves were normal. The coronary sinus (CS) was aneurysmally dilated (Fig. 2A and B, Video S1A and B), and in some views was reminiscent of a membrane partitioning the LA, closely mimicking a cor-triatriatum. Heavy spontaneous echo contrast (SEC) was visible in the hugely dilated RA, RV and dilated coronary sinus.

Supplementary video related to this article can be found at http://dx.doi.org/10.1016/j.ihj.2013.08.024.

Injection of agitated saline from the left antecubital vein was performed to rule out PLSVC draining to the CS. This resulted in opacification of the CS earlier than the main RA activity, confirming the presence of PLSVC draining to the CS. Injection of agitated saline into right antecubital vein also resulted in early appearance of the saline contrast in the CS prior to its appearance in the main RA cavity (Fig. 3, Video S2). Bilateral upper limb venogram demonstrated absent right SVC, presence of a bridging vein draining the right jugular and right subclavian veins which joined the left brachiocephalic vein to form the PLSVC (Fig. 4A, Video S3). The PLSVC was connected to the RA via a hugely dilated CS (Fig. 4B). He was treated with diuretics, digoxin, and amiodarone. At 3 months follow-up, he was in sinus rhythm which was maintained at 6 months follow-up. However, there was partial improvement in heart failure symptoms. The echocardiographic features of dilated RA, RV and coronary sinus and RV dysfunction remained same despite conversion into sinus rhythm.

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3. Discussion

We report an unusual case of isolated persistent left SVC with aneurysmally dilated RA, moderate RV systolic dysfunction, with persistent atrial tachycardia. This was associated with a hugely dilated CS which in the apical 4-chamber and parasternal long axis closely mimicked a cor-triatriatum. Saline contrast echocardiography and bilateral upper limb venogram confirmed the presence of an isolated PLSVC draining into the hugely dilated CS.

Persistent left SVC is one of the most frequent anomalies of the systemic venous circulation, occurring in 0.3–0.5% of the general population and in 4.3% of those with congenital heart disease.1–3 In most patients with persistent left SVC, a right SVC is also present, and both the vena cava drain normally into the right atrium through a dilated coronary sinus.4,5 Less commonly, the PLSVC may drain into the LA, producing a right-to-left shunt.

Persistent left SVC with absent right SVC (isolated PLSVC) as seen in our case, is a more rare entity and usually
associated with congenital heart disease and alterations of the cardiac situs.\textsuperscript{6–8} Contrary to most reported cases of isolated PLSVC, our case was unusual in the fact that there was situs solitus and no associated congenital heart disease, apart from the massively dilated RA with moderate RV systolic dysfunction. Hugely dilated RA, RV and coronary sinus in the absence of pulmonary hypertension or pulmonary stenosis and normal left sided chambers could be because of unexplained isolated RV cardiomyopathy.

Most cases of PLSVC with drainage to CS are asymptomatic and often discovered incidentally during surgery,
However, a higher incidence of cardiac arrhythmias and conduction defects has been reported in patients with PLSVC. This may be secondary to stretching and fibrosis of the atrioventricular node or His bundle by the dilated CS or associated sinus node dysfunction.9,10 Our patient also had a persistent atrial tachycardia which was perhaps due to the hugely dilated RA.

The markedly dilated CS in cases of PLSVC can often mimic clinical and echocardiographic features of supramitral ring or cor-triatriatum; the dilated CS has been known to even produce left ventricular inflow obstruction and heart failure.11,12 Presence of a dilated CS on echocardiography should always be considered as a clue to the presence of PLSVC which should be confirmed by saline contrast echocardiography and injection should be done separately through right and left antecubital vein to rule out associated absent right SVC. In our case, injection of agitated saline from the left and right upper arm veins resulted opacification of the CS before the RA. The final confirmation of isolated PLSVC was obtained following a bilateral upper limb venogram which confirmed absent right SVC, a bridging vein connection to the PLSVC, which then drained to the CS causing it to be aneurysmally dilated.

4. Conclusion

Presence of a dilated coronary sinus on echocardiography should alert the cardiologists to the presence of PLSVC. Agitated saline contrast echocardiography with injections into the bilateral antecubital veins should be important to establish the diagnosis and demonstrate the rare association of absent right SVC. It is also important to remember that occasionally the dilated CS in such cases may closely mimic the echocardiographic appearance of cor-triatriatum.

Authors’ contribution

Nagaraja Moorthy: Work-up of case, literature search.
Aditya Kapoor: Manuscript write-up, guidance.
Sudeep Kumar: Literature review, review.

Conflicts of interest

All authors have none to declare.

REFERENCES