Unroofed coronary sinus

Clinical Case Portal

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Authors:
Hoole SP, Cooper JC, Rusk RA
Papworth Hospital, Papworth Everard, Cambridge, Cambridgeshire, CBE 8RE

Case Report

The diagnosis of unroofed coronary sinus should be considered when investigating patients with possible intra-cardiac shunts causing right heart failure. Imaging with cardiac magnetic resonance and transoesophageal echocardiography made the pre-operative diagnosis possible, and clearly demonstrated the defect anatomy and blood flow through the defect on colour Doppler, respectively.

Patient history prior to current observation :
A 59 year old man presented with a 6 month history of worsening breathlessness, palpitations and peripheral oedema.

Clinical findings on admission, evolution and outcome :
Clinically he had fast atrial fibrillation and signs consistent with right heart failure. Initial transthoracic echocardiography showed a dilated, volume loaded right heart with severe tricuspid regurgitation. Cardiac catheterisation demonstrated elevated pulmonary artery pressures and a large left to right shunt at the level of the right atrium. Cardiac magnetic resonance (cardiac MR) imaging demonstrated a defect between the coronary sinus and left atrium (fig. 1). Subsequent imaging with transoesophageal echocardiography confirmed that this defect was a partially unroofed coronary sinus (fig. 2). Injection of
agitated saline via the left antecubital vein demonstrated normal right subclavian vein anatomy; opacification did not occur in the coronary sinus before the right atrium. The patient improved clinically with diuretics and anti-arrhythmic therapy and is due to undergo corrective surgery in the near future.

**Conclusion**

Unroofed coronary sinus is a rare cardiac anomaly in which there is communication between the coronary sinus and left atrium. This cardiac defect is strongly associated with left superior vena cava (LSVC) which commonly drains directly into the coronary sinus. The morphological type can be classified into four groups: type I, completely unroofed with LSVC; type II, completely unroofed without LSVC; type III, partial unroofed mid portion and type IV, partial unroofed terminal portion as in our case. It is important to consider this diagnosis when investigating patients with possible intra-cardiac shunts, as surgical correction with a baffle repair is curative. Imaging with cardiac MR (fig. 1) and transoesophageal echocardiography (fig. 2) has made pre-operative diagnosis possible.

**References**

1 - Ootaki Y et. al. Unroofed coronary sinus syndrome: diagnosis, classification and surgical treatment. Journal of Thoracic and Cardiovascular Surgery 2003: 126 (5); 1655-1656

Fig. 1 : Unroofed coronary sinus - Cardiac MR

Fig. 2 : Unroofed coronary sinus - TOE - Doppler colour flow