Unusual right heart failure due to a fistula between the circumflex coronary artery and the coronary sinus.

Clinical Case Portal

Date of publication:
24 Jul 2007

Topics: Heart Failure (HF)

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Case Report

This report illustrates the case of a 82 years old woman with heart failure secondary to a fistula between the circumflex coronary artery and the coronary sinus, and compares the ability of different techniques to assess the severity of a left to right shunt.

Patient history prior to current observation :

A 82 years old woman with a past history of an aortic valve replacement and CABG (LIMA-LAD) in 2001, came in our emergency room for a progressive shortness of breath starting a few weeks ago.
Clinical findings on admission, evolution and outcome:

Clinical examination revealed irregular heart beats, continuous cardiac murmur, oedema of legs and a jugular turgescence. Atrial fibrillation with an incomplete right bundle branch block was present on EKG. Blood analysis revealed hyponatremia and a elevated creatinin and urea. A transthoracic echocardiogram was performed to clarify the etiology of the dyspnea. A right ventricular enlargement was clearly identified fig. 1. The most surprising finding of this exam was the presence of an abnormal turbulent flow in the expected place of the coronary sinus. This striking flow ended in the right atrium fig. 2 and was systolo-diastolic as identified by the color M-Mode fig. 3. A transoesophageal echocardiography confirmed the presence of a severe dilation of the right ventricle fig. 4 and a significant tricuspid regurgitation fig. 5. At 45 degree, the left main coronary artery appeared dilated and a continuous turbulent flow was clearly identified inside fig. 6. At 90 degree, the circumflex coronary artery was also abnormally dilated fig. 7. During this exam, we have tried to follow the course of the flow, the circumflex coronary artery was in continuity with a bigger cavity, probably the coronary sinus. Magnetic resonance imaging was achieved to confirm this diagnosis. The fistula between the circumflex coronary artery and the coronary sinus was clearly demonstrated fig. 8 while the phase contrast imaging modality allowed us to quantify the shunt between the coronary artery and the right cavity with a ratio of 1.3. Finally, coronary angiography was performed. Selective opacification of the left coronary artery confirmed the dilation of the left main artery and dilation of the circumflex fig. 9. A large shunt is demonstrated between the circumflex and the coronary sinus with the opacification of the right atrium and the right ventricle. At the time of the catheterization, pulmonary hypertension persisted and a shunt ratio of 1.4 was measured.

Discussion

To explain the symptoms of our patient, we concluded that the congestive heart failure was multifactorial: a diastolic dysfunction due to atrial fibrillation but also and above all a chronic volume overload explained by the coronary fistula and the severe tricuspid regurgitation.

We have chosen to treat the patient by diuretic and resinusalization. In view of the age of the patient, a surgical approach of the fistula was not considered.

Conclusion

Two original points of this case report: first, the age of the patient and second, the fact that the circumflex was involved with the fistula which is rarely the case.

Video 1:

fistula between the circumflex coronary artery and the coronary sinus - TTE - right heart enlargement

Video 2:

fistula between the circumflex coronary artery and the coronary sinus - Turbulent flow in the coronary sinus
Fig. 1:
*fistula between the circumflex coronary artery and the coronary sinus* - MMode showing a systo- 
diastolic flow in the coronary sinus

![Image 1](image1.png)

Fig. 2:
*fistula between the circumflex coronary artery and the coronary sinus* - TEE - right ventricle dilation

![Image 2](image2.png)

Fig. 3:
*fistula between the circumflex coronary artery and the coronary sinus* - TEE - tricuspid insufficiency

![Image 3](image3.png)

Video 3:
*fistula between the circumflex coronary artery and the coronary sinus* - TEE - 45° - Color Doppler - 
Dilatation of the left main coronary artery with a turbulent flow inside

![Video 3](video3.png)

Video 4:
*fistula between the circumflex coronary artery and the coronary sinus* - TEE - Color Doppler - Dilation of 
the circumflex coronary artery with a turbulent flow.

![Video 4](video4.png)
Video 5:
*fistula between the circumflex coronary artery and the coronary sinus* - MRI - Fistula between the circumflex coronary artery and the coronary sinus

Video 6:
*fistula between the circumflex coronary artery and the coronary sinus* - Coronary angiography - Mega circumflex artery - Fistula between the circumflex and the coronary sinus