QUADRICUSPID AORTIC VALVE ASSOCIATED WITH AGENESIS OF CIRCUMFLEX CORONARY: A RARE CONGENITAL CARDIAC ENTITY TO SEE AT LEAST ONCE IN LIFE

Ilham BENSAHI1, 2, Amina ELFHAL1, 2, Raluca DULGHERU1, Yilmaz GORUR3, Luc PIERARD1, Patrizio LANCELLOTTI1

1. Department of Cardiology, Clinic of the valve, CHU Sart Tilman, Liege, Belgium
2. Department of Cardiology and Vascular Disease, CHU Ibn Rochd, Casablanca, Morocco
3. Department of Radiology, CHU de Sart Tilman, Liege, Belgium.

Introduction

Quadricuspid aortic valve is a rare malformation. Its incidence is estimated of 0.003 to 0.043% of all congenital heart disease (1, 2). It may be associated with other malformations (the most common being coronary artery anomalies found in around 10% of cases (3, 4)).

The case

We report the case of a 52 years old male patient presenting with new onset heart failure symptoms in the context of a dilated cardiomyopathy, in which transthoracic echocardiography (TTE) revealed a quadricuspid aortic valve (QAV), with no signs of stenosis and only mild to moderate centrovalvular regurgitation. 2D TTE raise the suspicion of a QAV and 2D and 3D transoesophageal echocardiography confirmed the presence of 4 equal-sized aortic cusps (figure 1). There was no hemodynamically significant impairment of leaflet opening (figure 2).

Coronary angiogram showed an occlusion of the distal right coronary artery. In addition, an aberrant coronary branch emerging from the left main was discovered (figure 3). The CT scan with 3D reconstruction provided clear images of the QAV, clarified the origin of each coronary ostia (figure 4): the left from the posterolateral left coronary sinus, and the right from the anterolateral left coronary sinus, and described perfectly the peculiar coronary anatomy of this case: agenesis of the circumflex coronary, presence of an aberrant branch emerging from the “left main” and heading towards the left atrium, and presence of a big “ramus intermedius” sharing the same origin with the LAD (figure 3). Chronic right coronary occlusion was confirmed.

Conclusion

Diagnostic of QAV is rare. Fortuitous discovery of this anatomic entity by TTE should trigger, besides assessment of valve hemodynamic, the search of other cardiac malformations such as coronary artery anomalies. The CT scan can provide a noninvasive accurate assessment of valve morphology and of coronary anatomy.

References