Double heart anomalies: left side accessory pathway associated to multiple coronary-pulmonary fistulae. A case study

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This case study describes the history of a 36-years-old healthy athlete who showed a normal resting ECG but during the warm-up revealed a typical postero-septal accessory pathway such as Left ventricular pre-excitation (Figure 1) which disappeared during the exercise test in the absence of symptoms and other abnormalities. The physical examination was normal and the family history was unremarkable for heart disease. In order to exclude the underlying cardiac diseases suggested by the Italian sports cardiology protocol (COCIS 2009), the athlete was subjected to a 2-D transthoracic echocardiography. This examination showed cardiac chambers of normal size and morphology with a conserved global and segmental kinetics, also heart valves were normal and well-functioning. However the color-Doppler examination in PSAX view revealed an anomalous double color flow jet in diastole arising from the lateral wall into the main pulmonary artery, and coronary artery fistula with non-significant left-to-right shunt (Qp/Qs ratio 1.2) came under suspicion (Figure 2) in the absence of signs of pulmonary and systemic overload. Consequently chest-cardiac computed tomography (CT) was performed, showing a complex anatomy of saccular dilated fistula that originates from all the proximal coronaries, more circumflex coronary artery, and drainages the main pulmonary artery was showed in detail by a 64 slice MDCT scanning. In particular the chest CT angiography showed laterally to the left of the proximal pulmonary artery trunk highlights the presence of a huge vascular malformation about the size of 17x17x9mm represented by multiple fistulous communications between arteries coronary-bronchial arteries and the pulmonary artery where proximity of the fistula is a greater apparent mediastinal arterial vascular plexus (Figure 3). A subsequent coronary angiography (Figure 4) confirmed the presence of numerous AV fistulas with coronary-pulmonary Left to Right moderate shunt as well as another communication between the descending aorta and a branch of the pulmonary artery with epicardial coronary disease-free. In view of the lack of symptoms and signs of ventricular overload, the athlete was considered eligible for competitive sport but require to be monitored with ECG + echocardiography every 6 months.