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Large Ostium Primum Interatrial Septum Defect in Asymptomatic Adult Athlete with Mitral Valve Prolapse and New Onset of Atrial Fibrillation

Colella A*; Crescenzi C; Tatangelo M; Calò L

Division of Cardiology, Policlinico Casilino, Via Casilina 1049, 00169 Rome, Italy.

Corresponding Author: Alessandra Colella

Division of Cardiology, Policlinico Casilino, Via Casilina 1049, 00169 Rome, Italy.

Email: alecolels@gmail.com

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Abstract

A large spectrum of congenital cardiac malformations can be the consequence of deficient formation of the embryonic atrioventricular canal by the endocardial cushions [1]. Among them, together with bicuspid aortic valve and mitral valve prolapse (MVP) [2], atrial septal defects (ASD) are one of the most frequent congenital heart diseases in adulthood. Differently from patients with secundum defects, ostium primum ASD are rarely reported in adulthood because those patients often require medical attention in pediatric age due to severity of symptoms. In ostium primum defect, in addition to the septal defect, the atrioventricular valves are often abnormal, including a cleft in the anterior mitral leaflet [3]. MVP is found in 30%-35% of ASD patients with or without significant mitral regurgitation (MR)and seems to be secondary to an abnormal left ventricular geometry and dilatated right ventricle which leads to shortening of inter-papillary distance and redundant chordae tendineae [4]. Atrial fibrillation (AF) is the most common cause of hospital admission among all arrhythmias in general population and AF is also the most common arrhythmia in the athletic population [5].

We report the case of an adult athlete with recent onset of asymptomatic AF discovered during yearly pre-participation screening n whom, after an unsuccessful electrical cardioversion, noninvasive and invasive exams clarified the diagnosis of ostium primum defect and severe MVP; the patient was then referred to a specialized surgical center.

Keywords: Congenital heart disease; Sports cardiology; Atrial septal defects; Echocardiography; Mitral valve prolapse; Ostium primum; Atrial fibrillation; Athlete.

Case report

In a 47-year-old male patient (height 184 cm; body weight 72 kg), professional martial arts instructor always eligible for competitive sport, was reported asymptomatic new onset AF during pre-participation screening. His previous cardiological history consisted in MVP with significant MR but nor symptoms nor limitations of his sports activity. Further electrocardiograms (EKGs) were performed and showed persistent AF after 6 months. Thus, he was admitted to our hospital for electrical cardioversion (EC) after premedication with amiodarone and DOAC. EKG on admission showed AF with ventricular rate response of 60 beats/min and in complete right bundle branch block; blood tests were in the normal range except mild TSH increase. Proper sinus rhythm was restored after 3 shocks and intravenous flecainide but 3:2/4:3 atrioventricular block at a frequency of 60 bpm occurred. The patient was asymptomatic and he remained under observation. Few hours later he had an arrhythmic recurrence of AF and disappearance of the atrioventricular block. The patient was then referred to the dedicated outpatient clinic where he was re-evaluated after two weeks. EKG was once again performed and it showed AF with ventricular rate response of 67 beats/min, incomplete right bundle branch block and nonspecific repolarization abnormalities. Physical examination revealed normal blood pressure, 3/6 L sys-

tolic murmur over all areas, no signs of pulmonary congestion. A new two-dimensional transthoracic echocardiography (TTE) confirmed the presence of the already known MVP with severe mitral regurgitation (PISA 11 mm, V 36 cm/s) secondary to MVP and showed significant left atrial enlargement (LAVI 62 ml/m²) together with mild right atrial enlargement. Left ventricle was mildly hypertrophic with normal size and normal ejection fraction. For further evaluation, to define better the entity of valvulopathy and its possible arrhythmogenic burden, a transesophageal echocardiography (TEE) and 24 Hour EKG Holter Monitoring were performed. TEE revealed a large ostium primum type ASD (43 x 45 mm) with severe left-to-right shunt Qp/Qs=3. Mitral valve appeared dysmorphic, with a cleft of mitral valve posterior leaflet P2-P3 and multiples jet along all the commissure determining severe regurgitation (V 48 cm/s). The exam also confirmed biatrial enlargement and showed significant right ventricular dilatation (RVD1 57 mm, RVD2 55 mm apex-to-base length 82 mm) with conserved tricuspid annular plane systolic excursion (TAPSE). 24 Hour EKG Holter Monitoring registered AF with coarse fibrillatory waves with an average ventricular rate response of 62 beats/min for the whole time. Multiple episodes of non-sustained ventricular tachycardia (NSVT) were registered, the longest consisted in 18 s, all of them without symptoms reported in patient's diary. Antiarrhythmic therapy with nadolol was then prescribed. In addition, the patient underwent to cardiac magnetic resonance (CMR) which confirmed the presence of ASD with severe left-to-right shunt (Qp/Qs 3.3) and bileaflet MVP. CMR also described the presence of mitral annular disjunction (MAD)of 6 mm, moderate left ventricular and severe right ventricular dilatation (EDV 219 mL/m²) both with preserved function. Neither LGE nor myocardial edema in T2-weighted imaging were reported. The patient was referred to a center specialized in cardiac surgery for the defect's closure and mitral valve's management.

Discussion

This case is relevant because of the age of the patient, coexistence of different cardiac conditions, lack of symptoms and normal tolerance to exercise. It is true that an isolated ASD could be undiagnosed for years and be occasionally discovered in adulthood, but frequently it's about smaller ostium secundum ASD. Ostium primum defects are instead infrequent findings in adults because usually they're associated with AV valves impairment and haemo-dynamically significant shunt leading to cardiac surgery in younger age. In literature few cases of asymptomatic adults with ostium primum are described, but none of them regarding athletes. Our patientis a 47 years old man with ostium primum like defect in association with MVP determining severe MR. MVP is highly prevalent in patients with ASD, apparently due to the presence of an organic primary process of the MV apparatus and an abnormal left ventricular geometry and dilated right ventricle. MVP is the most frequent cause of organic MR in Western countries and by itself, when causes from moderate to severe MR, may produce hemo-dynamically significant effects. Often patients experience fatigue, dyspnea, exercise intolerance, palpitations which require medical attention. Our patient's heart has the characteristic remodeling found in left-to-right shunt consisting in impaired right atrial reservoir, right ventricular dilatation and pulmonary-to-systemic flow ratio exceeding 1.5, which triggers a cascade of changes in the myocardium and in the pulmonary vasculature. This remodeling coexists with changes produced by MVP involving principally left atrium and ventricle. It appears surprising his tolerance to exercise, considering his athletic history and daily activities as martial arts instructor. According to ESC 2020 guidelines, martial arts is considered a medium intensity power discipline [6] where repeated bursts determine a substantial increase in heart rate and blood pressure. Long-timepractice may lead to an increase in left ventricular wall thickness and modest increase in left ventricular cavity size and function as well. The patient suffers also from arrhythmias that are known to be associated with both ASD and MVP. He underwent pre-participation screening every year, including exercise testing with normal exercise test response, but AF or NSVT were not founding previous medical examinations. It's likely that is a new onset AF which depicts the evolution of the pathology. Our case highlights the importance of cardiovascular disease prevention even in apparently healthy adult athletes.

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