Association between Ebstein Anomaly and Coronary Artery Disease: A Report Case

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Abstract: The intention of this article is to report a case of a 26 years old man presenting a rare combination of congenital heart disease (Ebstein anomaly) and coronary artery disease. EA (Ebstein anomaly) has a prevalence of 1% of all congenital heart diseases, as CAD (coronary artery disease) has a little evidence in literature’s registries when occurred in individuals with 40 years old or less. Therefore, this case report rises attention to the rarity of those patologies, which individually are already considerably rare and, in this case, it comes associated what turns this diagnosis exceptionnal, highlighting the complexity of the treatment.

Key words: Ebstein, infarction, young.

1. Introduction

CAD (coronary artery disease) is the leading cause of deaths globally [1] and remains a huge burden to health systems over the world. Because of its prevalence, the disease pathophysiology has been extensively studied. The known cardiovascular risk factors has a population attributable risk higher than 90% for the myocardial infarction [2] and age plays a central role in the coronary obstructions, as atherosclerosis mechanisms requires years of action [3]. Young patients presenting CAD commonly have strong association with risk factors, as familial history of premature CAD, obesity [4] or heavy smoking [5]. Additionally, in approximately 20% of young patients with clinical presentation of myocardial infarction a non-atherosclerotic coronary disease have been diagnosed [6], such as coronary anatomy abnormalities, vasoconstriction [7] or thrombophilia due to autoimmune diseases. Cocaine and other illicit drug use have been increasingly associated with acute myocardial infarction [8] and accelerated atherosclerosis [9].

Despite all knowledge accumulated over the decades about atherosclerosis and CAD, the causes of the disease in some cases are still uncertain, because they may not be clearly associated to any known cardiovascular risk factor, even after intensive investigation, or have strong association with a disease that had never been previously related with CAD.

Ebstein anomaly is a rare syndrome characterized by the ventricular implantation of tricuspid valve, leading to right atrium enlargement, right ventricle failure [10] and, when it is diagnosed during early stages of life, death occurs commonly in childhood [11]. Because of that, no evidence of association between Ebstein Syndrome and CAD has been found in the literature. We report a young adult with Ebstein anomaly presenting with severe CAD due to atherosclerotic coronary artery disease without traditional cardiovascular risk factors.

2. Methods

The information of this case report was collected by medical records revision, patient’s anamnesis, physical examination, diagnostics methods and literature revision. The patient provided written informed consent for the case report.
3. Case Report

A 26 years old male was admitted to emergency unit with a history of a recent onset of intense constrictive chest pain associated with sweating. ECG and qualitative troponin measurement on admission evidenced NSTEMI (non-ST elevation myocardial infarction). Immediate transfer to a coronary care unit was requested. The patient referred fatigue three months before hospitalization although no medical attention was sought in that period. There was no history of hypertension, diabetes, hypercholesterolemia, illicit drugs or tobacco use, as well as familial premature coronary disease or sudden death. At physical examination, there were systolic mitral and tricuspid murmurs and BMI (body mass index) was 21.6 Kg/m². The coronary care unit admission ECG showed Q waves at V1-V3 and T wave inversion at DII, DIII and aVF leads and troponin level 0.906 ng/mL (ULN 0.04 ng/mL). An transthoracic echocardiogram demonstrated downward tricuspid valve displacement, tricuspid regurgitation, right atrium enlargement, and right ventricular dysfunction, characterizing Ebstein’s syndrome (as shown in Fig. 1). Mild left ventricular systolic dysfunction was also observed.

A coronary angiogram was performed 48 h after admission in the coronary care unit and demonstrated proximal severe obstruction followed by occlusion of left anterior descending artery in its middle third (Fig. 2), subocclusive plaque in proximal right coronary artery and severe obstruction in proximal portion of posterior ventricular artery (Fig. 3). The left main and circumflex arteries were free of obstruction.

As the symptoms began more than 96 h before the angiography, patient was submitted to percutaneous coronary angioplasty and two bare metal stents were implanted in the right coronary and the posterior ventricle arteries, respectively (Fig. 4). The approach to left anterior descending artery occlusion was postponed to the heart team meeting.

Approximately 20 days after successful angioplasty, patient was submitted to a cardiac MRI that demonstrated a 20 mm/m² displacement of tricuspid valve septal leaflet towards to the apex cordis and reduced functional right ventricle volume with atrialized region above tricuspid valve ring, confirming Ebstein’s Anomaly. Additionally, there was a subendocardial pattern late gadolinium enhancement at apical and basal-inferior mid-inferior, mid-anterior, apical-anterior and septal walls, in agreement with the coronary obstructions in angiography, characterizing myocardial infarction in

Fig. 1  Admission echocardiography.
Fig. 2  Coronary angiogram: demonstrating proximal severe obstruction followed by occlusion of left anterior descending artery in its middle third.

Fig. 3  Coronary angiogram demonstrating suboclusive plaque in proximal right coronary artery and severe obstruction in proximal portion of posterior ventricular artery.
Fig. 4  Right coronary artery after bare metal stent implantation. Note complete resolution of coronary obstructions. The guide wire can be seen, extending to posterior ventricular artery.

Fig. 5  Cardiac resonance.
two regions and left ventricle infarcted mass was estimated at 12% (Figs. 5 and 6).

Laboratory investigation was performed to evaluate traditional coronary artery disease risk factors and Total Cholesterol was 139 mg/dL, LDL-Cholesterol 86 mg/dL, HDL-Cholesterol 43 mg/dL, Triglycerides 53 mg/dL, Glucose 80 mg/dL. The autoimmune diseases were discharged with negative tests for rheumatoid factor, antinuclear antibodies, p- and c-ANCA, Anti-SCL 70 and anti-Jo1 antibody. Serologies for A, B and C hepatitis, HIV and Chagas’ disease were also negatives.

Patient was treated with ASA 100 mg OD, clopidogrel 75 mg OD, simvastatin 40 mg OD, carvedilol 12.5 mg BID and ranitidine 150 mg BID. After the angioplasty, patient relieved the symptoms and myocardial necrosis markers tests were consistently negative. The ACE inhibitor was initiated as patient presented left ventricular dysfunction at echocardiogram, but it led to hypotension and renal function reduction and, because of that, it was interrupted. After heart team discussion, it was decided to perform combined surgical approach of Ebstein’s anomaly and the anterior descending artery occlusion in the same time, with two specialists surgical teams, one in each surgery, and the procedure will be performed further only if patient develop new or worsening symptoms of angina or heart failure. However, patient was discharged from the hospital after forty days and, during the 30-days follow-up in the outpatients’ clinics, he was clinically stable, without new onset of symptoms.

4. Discussion

The present case describes an atherosclerotic acute coronary syndrome in a young active adult presenting Ebstein’s Anomaly, not diagnosed during the early ages of life, without any complications during childhood and any related symptoms in the adult age. This is an unusual evolution of that rare disease and, as far as we are concerned, not previously described association with coronary atherosclerosis in the scientific literature.

The echocardiogram and cardiac MRI clearly evidenced the association of two different pathophysiologic processes: the apical displacement of the tricuspid valve and the definite regional ischemic injury of the left ventricle. Although some Ebstein’s Anomaly patients can present left ventricle dysfunction [12], the coronary angiogram confirmed...
the atherosclerotic origin of coronary obstructions in agreement with cardiac MRI findings. Pathophysiologic mechanisms of the association between Ebstein’s anomaly and atherosclerosis are not stated, but maybe Ebstein’s anomaly might accelerate the development of atherosclerosis. The mild abnormalities of Ebstein’s anomaly in this case might allow the development of atherosclerotic injuries that were not observed in other more severe patients. The absence of other cardiovascular risk factors also highlighted the association of both conditions.

Ebstein’s anomaly is a rare congenital disease, occurring in one to five on each 200,000 live births, and 1% prevalence of all congenital heart diseases [13]. Besides its prevalence, the prognostic is poor when the disease is diagnosed in first days or weeks of life, with 1- and 5-year survival rates estimates as 78.6% and 76.3%, respectively [14], most often requiring surgical treatment. Besides the right ventricle characteristics, Ebstein’s anomaly is commonly associated with other congenital defects, such as mitral valve prolapse and left ventricular noncompaction [15].

When Ebstein’s anomaly is diagnosed in the adulthood, the treatment is challenging. As patients have commonly mild valve disorders and the survival is high [16], the surgical treatment of associated with higher rates of need of new surgeries and, because of that, surgical procedures are indicated only when Ebstein’s anomaly is related to symptoms worsening. In patients with increased risk of arrhythmia, ablation procedures may be considered [16]. Our patient haven’t presented any associated defects described in the literature that we quoted above or any symptoms due to Ebstein’s anomaly, the condition was only diagnosed after acute coronary syndrome. The clinical presentation has been occurred due to acute coronary syndrome and relieved after percutaneous coronary interventions. Additionally, anterior descending artery occlusion was considered as a chronic condition, in which revascularization would be associated with major clinical event reductions only in patients with significant ventricular dysfunction [17, 18].

On the other side, CAD in younger ages is commonly associated to strong risk factors or familial history, which were ruled out in our young adult patient. As far as we are concerned, this is the first report of an association between EA and significant CAD atherosclerotic lesion in a young adult. In the present case, the definitive treatment includes coronary artery and the tricuspid valve surgical interventions. The planned approach consists in a repair that comprises the reconstruction of a normally shaped right ventricle and the repositioning of the tricuspid valve at the normal level developed by Carpentier et al. [19] and a CABG (coronary artery bypass graft) at the same surgical time. We believe both procedures together might reduce the risk of it procedure. However, Ebstein’s anomaly correction and CABG combined procedures—and its risks—were not described previously in the literature. Therefore, our patient is being closely followed at outpatients’ clinics and surgical procedure will be indicated in case of symptoms or left ventricular function worsening.

5. Conclusions

All clinical facts that were discussed in this case report rises our attention for medical rarity and a new challenge rises to overcome. A literature review of these subjects was discussed aiming to clarify the treatment of this rare combination of cardiopathies. Therapy in this case is a challenge, since there is low evidence in literature about management of these patients, consequently, new studies should be carried out.

References


