

Case Report

Ebstein Anomaly in a 60 Years Patient: A Lucky Finding

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Abstract: Ebstein anomaly is a rare congenital disease of the tricuspid valve (<1%) diagnosed at all ages. A single case of an 85 y old patient was reported in 1979 as the longest survival with ebstein anomaly who had no cardiac symptoms until 79 years old. The aim of this case report is to highlight the need for an early echocardiographic diagnosis of this disease to prevent sudden death from arrhythmias or other complications because as we see patients with ebstein anomaly can live a healthy long life asymptomatic. The patient described in this case is a 60 years old male diabetic and heavy smoker, who presented to the cardiology department with fatigue and atypical angina with dyspnea on moderate effort. Cardiac ultrasound was in favor of an isolated Ebstein anomaly type A, with partial atrialization of the right ventricle (RV) with an adequate volume of the right ventricle (17cm²) and no specific other associated anomalies. Symptoms described by the patient were purely pulmonary due to a mild obstructive disease. Patient was diagnosed with chronic obstructive lung disease due to his smoking habits. Reaching this age and being asymptomatic with conserved RV and LV function is a sign of good outcome. This case was an interesting lucky finding. It was astonishing to see a patient surviving this anomaly at 60 years old asymptotically.

Keywords: Ebstein Anomaly, Asymptomatic, Atrialization

1. Introduction

Ebstein anomaly is a congenital disease of the tricuspid valve, associated with a displacement of septal and posterior tricuspid leaflets toward the apex of the right ventricle (RV). It results in an atrialization of the right ventricle. It's a rare abnormality diagnosed at all ages. The age at presentation depend on the presence of associated diseases and severity of the tricuspid regurgitation [1]. There is an increase in the prenatal diagnosis of this disease due to the development of this anomaly in fetal life. 1 in 20000 babies are diagnosed with ebstein anomaly (<1%). [1] It can be medically induced by benzodiazepines, antihypertensive drugs, valproic acid, marijuana, and organic solvents through pregnancy. The association with lithium was disputed. Some new emerging data about maternal β -thalassemia need to be confirmed [2].

A single case of an 85 y old patient was reported in 1979 as the longest survival with ebstein anomaly who had no

cardiac symptoms until 79 years old. Older age patients often present with palpitations due to arrhythmias or mild symptoms such as exertional dyspnea and sometimes fatigue. Myocardial fibrosis of the right ventricle due to this anomaly was a predictor of high rate of arrhythmias and even complicated by heart failure symptoms. The key to prevent arrhythmias and other complications is early diagnosis in adult patients with long term and good outcomes. [1]

2. Clinical Case

A 60 years old patient, with a history of diabetes mellitus 7 years ago, on glimepiride 4mg daily not very well controlled on treatment, dyslipidemia on simvastatin and a heavy smoker presented for chest pain. 1 month ago patient complained of fatigue and atypical angina. His pain was rarely oppressive not irradiating to his left arm neither associated with diaphoresis sometimes exacerbated with exertion.

Furthermore, the patient noticed some episodes of

intermittent dyspnea on moderate effort. He also complained of chronic fatigue and weight loss since 2 years. He had normal vital signs. On physical examination, S1 and S2 were normal with no associated murmurs. Lungs were clear but some bilateral basal wheezing. His chest x- ray revealed a normal heart size with overinflation and flattening of the diaphragm. Patient is in sinus rhythm with no ST-T changes, preexcitation or any other ECG modifications. Cardiac ultrasound was in favor of an isolated ebstein anomaly type A (Carpentier et al 1988), with partial atrialization of the right ventricle (RV) with an adequate volume of the right ventricle (17cm²) and no specific other associated anomalies (no RV infundibular obstruction, pulmonary atresia or shunts), but the interatrial septum is mildly aneurysmal. The rest of the echo revealed a normal LV function, normal strain (GLPS average=-18%), normal diastolic function with normal LVEDP. The right ventricle is not dilated. Tricuspid regurgitation is grade I associated to a Systolic pulmonary artery pressure (SPAP=16mmHg) and an anomalous motion of the interventricular septum. In addition, a tricuspid valve apical displacement >8mm/m² (in this case it's =17mm/m²) was pathognomonic in this anomaly [figure 1]. Cardiac catheterization was done to rule out any associated coronary anomalies. It revealed normal coronary arteries.

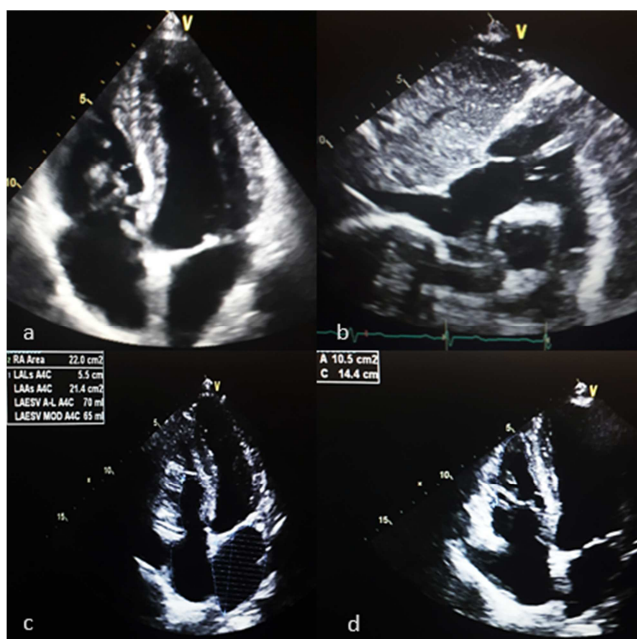


Figure 1. Cardiac ultrasound.

- 4 chambers view revealing an atrialization of the RV (Apical displacement of the septal leaflet of tricuspid valve =17mm/m²)
- Subcostal view
- 4 chambers view: measurements of the right and left atrium
- 4 chambers view: RV measurements

3. Discussion

The long term follow up of this anomaly is rare due to the low prevalence of the disease with high variation in its anatomic components.

In a retrospective review of 51 patients with ebstein anomaly, the mean age at diagnosis was 21 years +/-21. The overall survival in this single center study demonstrated 100% survival to 40 years, 95% to 50 years and 81% to the age of 60. [3].

In patients with mild Ebstein anomaly, survival was reported to the ninth decade. A single case of an 85 y old patient was reported in 1979 as the longest survival with ebstein anomaly who had no cardiac symptoms until 79 years old. These patients may be asymptomatic for a long time. Patients with Ebstein's anomaly who reach late adolescence and adulthood often had a long term outcome. In the other hand, fetal and neonatal presentations have a poor outcome. [4] Our patient was at 60 years old when he presented for a cardiac workup. Reaching this age and being asymptomatic with conserved RV and LV function is a sign of good outcome. Ebstein anomaly includes many features associating the adherence to the myocardium of the tricuspid valve leaflets with an apical displacement of the septal and posterior leaflets.

Atrialization of the right ventricle with different degrees, involving a redundant and fenestrated anterior leaflets. Various degree of the tricuspid regurgitation and cyanosis with different presentations. In addition, dilation of the right atrium could be also included.

Other echocardiographic anomalies could be affiliated to this anomaly as ASD or PFO, VSD, mitral valve prolapse, RVOT obstruction and LV anomalies. Electrophysiological anomalies as accessory pathways and atrial tachycardia may be involved and catheter ablation could be a solution [5, 6, 7]. Heart failure is also a severe consequence of ebstein anomaly. In this case report, the patient had no associated congenital malformation nor arrhythmias. [8]. A study on 37 patients with ebstein anomaly in adulthood aged 43.0 ± 14.4 years revealed that fibrosis was the initial cause of arrhythmias and heart failure assessed by LGE on cardiac MRI. 20% of patients reaching adulthood died from arrhythmias, whereas 50% died from heart failure [9]. Sudden cardiac death may occur in asymptomatic patients with mild disease and normal cardiothoracic index on chest-X-ray. There is no evidence for tricuspid surgery in reducing the incidence of sudden cardiac death in asymptomatic patients. Risk of surgery is high [10]. In this case, patient was treated symptomatically as there is no evidence to prove that survival will be better after surgery. In 1999, a case report revealed an association between ebstein anomalies with myocardial bridging and anomalous coronary artery [11]. As for this case a cardiac catheterization didn't reveal any bridging, his symptoms were purely pulmonary with mild obstructive pattern on PFTs. The standard diagnostic tool for this entity is the transthoracic echocardiography [12]. Magnetic resonance imaging (MRI) is not used routinely [8]. On the other hand, associating Cardiac MRI with transesophageal echocardiography and rarely cardiac catheterization provide additional information for surgical decision making [13]. Carpentier et al. classified the ebstein anomaly in 1988 in 4 types; type A with an adequate volume of the true RV, type B with large atrialized

RV but a freely moved anterior leaflet, type C associating an obstruction of the RVOT with a severely restricted anterior leaflet and type D embracing an almost complete atrialization of the RV. A prospective study on thirty –two patients with ebstein anomaly concluded that it seems that size of the RV depend on the degree of the tricuspid regurgitation what it look to be in the described case above [14]. Furthermore, dilatation of the right heart with decreased contractility are features of poor prognosis. The echocardiographic features for the diagnosis of this anomaly embrace: on M-mode, a paradoxical septal motion with delayed closure of tricuspid valve leaflets. On the other hand, on Two-dimensional echocardiography an apical displacement more than 8mm/ m² of the septal leaflet with eccentric leaflet coaptation. Same features were present in the echocardiography of our case and helped to put the diagnosis [8]. Let's also notice that the electrocardiogram of the patient was normal with no associated signs of delayed or prolonged duration of the RV which is a representative of a mild disease. [15]

4. Conclusion

In the end, we described a rare case of ebstein anomaly who presented at 60 years old for cardiologic assessment. Patient had some atypical chest pain with dyspnea on exertion that were mainly explained by an obstructive pattern because the patient was a heavy smoker. His echocardiography revealed an ebstein anomaly with conserved RV and LV function with no dilatation nor a significant tricuspid regurgitation. It was an interesting lucky finding. It was an astonishing case surviving this anomaly at 60 years old asymptotically. This case highlight the need for further preventive and early diagnosis strategies to prevent complications and sudden cardiac death in adults with ebstein anomalies as arrhythmias and congestive heart failure, because as we noticed it's a disease if early diagnosed and followed up carefully can remain asymptomatic for a long time with a very good prognosis in adults.

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