

A Rare Presentation with Angina and Pseudoinfarct ECG Pattern in A Patient with Severe Form Ebstein's Anomaly

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ABSTRACT

We are presenting a rare case of an elderly female who had been asymptomatic until the age of 56 years and had successfully completed three pregnancies. She presented with an episode of acute onset chest pain of moderate intensity and was initially suspected to have an evolved inferior wall myocardial infarction on the basis of the electrocardiogram. The echocardiogram performed subsequently surprisingly revealed the presence of a severe form of Ebstein's anomaly.

Various forms of electrocardiographic abnormalities and conduction defects are reported in Ebstein's anomaly but the presence of a normal ECG pattern or pseudoinfarct pattern with the absence of any associated conduction blocks or axis deviation is highly unlikely especially in severe forms of the disease. There are three aspects of this cases which make it rare. Firstly, the presentation of a patient with severe form of Ebstein's anomaly with an isolated evolved inferior wall myocardial infarction pattern on the ECG, secondly the presence of an extra-ordinarily long asymptomatic period and thirdly the presentation with angina as the presenting symptom with normal epicardial coronaries.

Keywords: Ebstein's, pseudoinfarct pattern, angina, inferior wall infarction pattern, normal coronaries

INTRODUCTION

Ebstein's anomaly is a rare congenital heart disorder occurring in 1 per 200,000 live births and accounting for 1% of all cases of congenital heart disease¹. This anomaly was described by Wilhelm Ebstein in 1866^{2,3,4}. The cardinal symptoms in Ebstein's anomaly are cyanosis, right-sided heart failure, arrhythmias, and sudden cardiac death. The hemodynamic variations and clinical presentation depend on the age at presentation, anatomic severity, hemodynamics, and degree of right-to-left inter-atrial shunting in patients with severe forms of Ebstein's anomaly adult sur-

vival is less common and most of the patients do present with heart failure or arrhythmic symptoms early in adulthood.

We are presenting a rare case of a previously undiagnosed elderly acyanotic female who had tolerated three pregnancies successfully and had been apparently asymptomatic till the age of 56 years when she presented with an episode of acute onset typical chest pain of moderate intensity and was initially suspected to have an evolved/old inferior wall Q wave myocardial infarction pattern on the basis of the electrocardiogram while the echocardiogram subsequently revealed the presence of a severe form of Ebstein's anomaly and the coronary angiogram was normal.

Case Description

A previously undiagnosed elderly female with the history of chest pain of moderate intensity which was typical for angina presented to the emergency room and the

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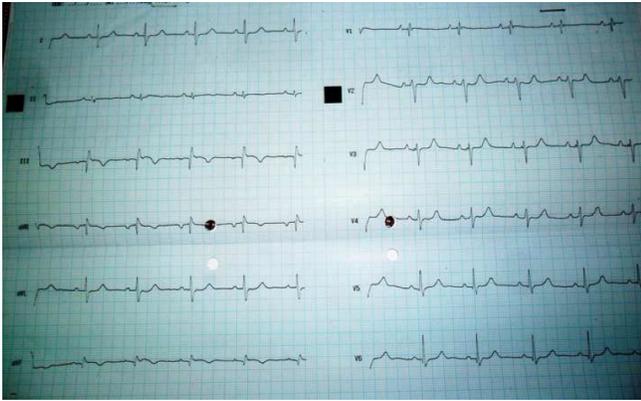


Figure 1: ECG of the patient showing q waves in leads II, III, aVF resembling an evolved/old inferior wall myocardial infarction pattern

electrocardiogram (Figure 1) revealed a pattern suggestive of an old inferior wall myocardial infarction which was initially suspected in her but surprisingly the echo cardiogram revealed that the patient had a severe form of Ebstein's anomaly with the left ventricle entirely compressed by the right and the presence of an atrial septal defect which was shunting left to right. (Figure 2–4, Video 1–3)

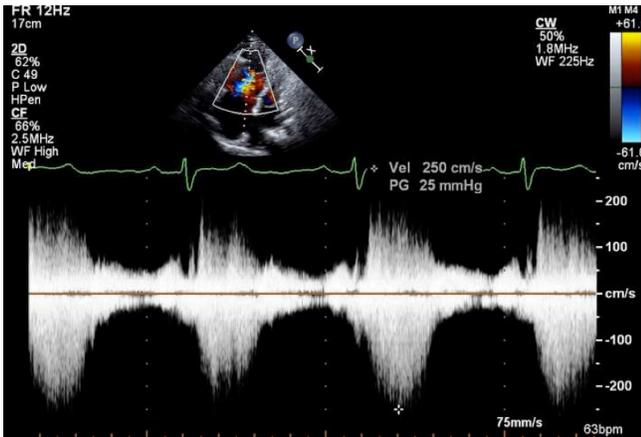


Figure 2: Continuous wave Doppler signal across the tricuspid valve showing low pressure tricuspid regurgitation of severe intensity

The patient had no prior history of chest pain or any symptoms of exertional dyspnoea and was able to perform all her routine activities and her daily chores with the presence of mild fatigue and dyspnoea on unaccustomed activity which she considered normal for her age. There was no prior history of chest pain or exertional angina. The echocardiogram also did not reveal any regional wall motion abnormality in the inferior wall territory which would suggest the presence of an infarct and the cardiac enzymes were also normal. The patient was taken up for a diagnostic coronary angiogram



Figure 3: Colour Doppler still across the atrial septal defect showing a left to right shunt

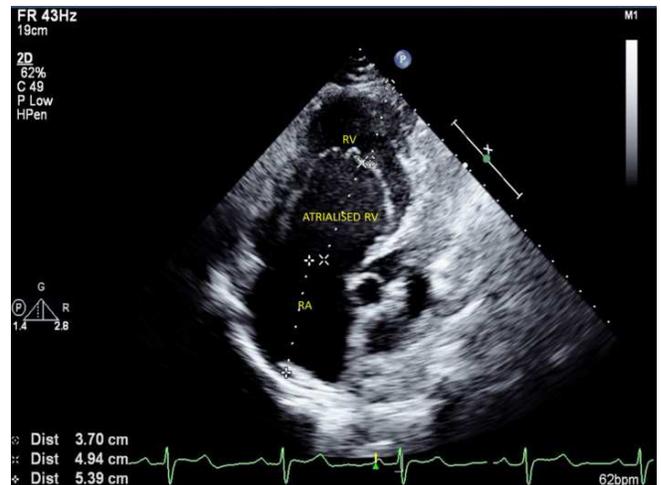


Figure 4: Measurement of the right atrium, right ventricle and the atrialized right ventricle

which turned out to be normal. The X-ray showed the presence of cardiomegaly with the presence of dilatation of right sided chambers. (Figure 5)

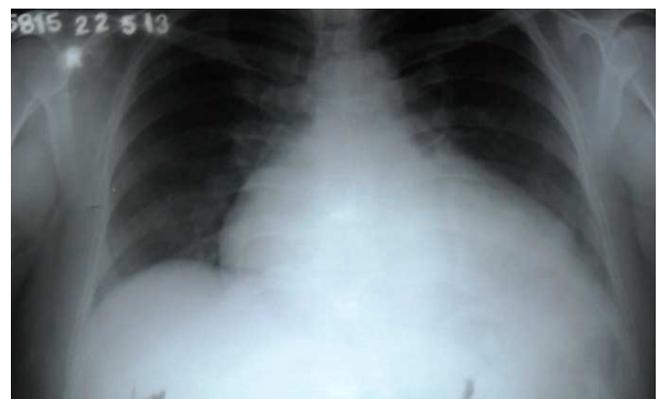


Figure 5: X-Ray of the patient showing gross cardiomegaly with dilated right sided chambers

The anginal pain was subsequently thought to be due to strain on the right and the left ventricles or possibly due to micro vascular angina although the epicardial vessels and the blush grade were normal during the angiogram. The cause of the abnormal ECG pattern was considered to be an intra-ventricular conduction abnormality of an atypical form which made the ECG to appear in a pattern akin to an evolved or old inferior wall myocardial infarction.

The saturation was 92% at room air and the patient was acyanotic which again is extremely uncommon in elderly patients having severe form of Ebstein's anomaly.

The patient was subsequently taken up for an elective repair of the tricuspid valve with reduction of right atrial size. The gross morphological of the left ventricle suggested a normal left ventricle with no evidence of an infarct. Histopathological assessment was not done as left ventricular endocardial biopsy was not done as it was not clinically indicated and it would have increased the surgical risk and subsequent risk of arrhythmias. The post-surgical ECG was similar to the previous and the pseudo infarct was persistent. The surgical outcome was good at 2 months of follow up the patient was maintaining adequate functional capacity and has NYHA class II symptoms. The patient has been subsequently lost to follow up.

DISCUSSION

Ebstein's anomaly is a complex congenital anomaly with a broad anatomic and clinical spectrum. Management is complex and must be individualized. Precise knowledge about the different anatomic and hemodynamic variables, associated malformations, and management options is essential.

This case is an interesting experience to understand the varied presentation of Ebstein's anomaly which is most often a disease which presents in infancy or in early childhood. This case forms a valuable learning opportunity for clinicians and primary care physicians dealing with adult patients having coronary artery disease or various systemic illnesses which can have varied presentation and can present with chest pain and similar symptoms. Ebstein's anomaly can have various forms of electrocardiographic abnormalities and can have various forms of conduction anomalies but presenting with an isolated pseudoinfarct pattern with the absence of any axis deviation and even the absence of any degree of

cyanosis with a left to right shunt across the ASD is an extremely unlikely although a possible presentation.

Ebstein's anomaly is commonly diagnosed during fetal echocardiography because in severe cases there may be cardiomegaly, hydrops, and tachyarrhythmias^{5,6}. Neonates with Ebstein's anomaly may present with cyanosis, congestive heart failure and marked cardiomegaly⁵. Symptomatic children with Ebstein's anomaly may have progressive right sided heart failure, but adult survival is common although patients being asymptomatic for extremely long periods is extremely unlikely in patients having a severe form of the disease.

Children more than 10 years of age and adults often present with arrhythmias⁷. Adults also present with progressive cyanosis, decreasing exercise tolerance, fatigue, or right-sided heart failure. In the presence of an interatrial communication, the risk of paradoxical embolization, brain abscess, and sudden death increases⁸. Exercise tolerance is dependent on heart size and oxygen saturation^{8,9}.

The ECG is abnormal in most patients with Ebstein's anomaly. It may show tall and broad P waves with complete or incomplete right bundle-branch block patterns¹⁰ and bizarre morphologies of the terminal QRS pattern result from infra-Hisian conduction disturbance and abnormal activation of the atrialized right ventricle¹¹. Complete heart block is rare but first-degree atrio-ventricular block occurs in 42%^{10,12}.

From 6% to 36% of patients with Ebstein's anomaly have more than one accessory pathway^{10,11}, and most accessory pathways are located around the orifice of the tricuspid valve¹².

The cardiac silhouette may vary from almost normal to the typical Ebstein's anomaly configuration consisting of a globe-shaped heart with a narrow waist similar to that seen with pericardial effusion. Vascularity of the pulmonary fields is either normal or decreased. A cardio thoracic ratio >0.65 carries a poor prognosis. This patient had gross cardiomegaly but the patient was still minimally symptomatic. Diagnostic cardiac catheterization is rarely necessary in patients with Ebstein's anomaly, other than for pre operative coronary angiography. Right ventricular and pulmonary artery pressures are usually normal in patients with the anomaly, although the right ventricular end-diastolic pressure may be increased.

Several studies have reported on the natural history of Ebstein's anomaly^{13,14}. The largest of these studies, report-

ing on 505 patients with Ebstein's anomaly, was published more than 30 years ago in the pre-echocardiographic era¹⁴. Of the infants presenting before 1 year of age, 72% were in heart failure, but for 81% of the others, growth and development during infancy were average or good and 71% of children and adolescents and 60% of adults were in NYHA functional class I or II¹⁴.

Of those who had surgical treatment, 54% did not survive the operation.

Celermajer et al¹⁵ reviewed 220 cases of Ebstein's anomaly with 1 to 34 years of follow-up. Actuarial survival for alive-born patients was 67% at 1 year and 59% at 10 years. Predictors of death were echocardiographic grade of severity at presentation (relative risk increased by 2.7 for each increase in grade), fetal presentation, and right ventricular outflow tract obstruction. In rare cases, patients with Ebstein's anomaly can rarely live upto 70 years, but 1 reported patient died at 85 years of age^{16,17}. A reassessment of the prognosis of Ebstein's anomaly is appropriate in the current era of refined cardiovascular intervention.

The most amazing feature of this case were that the patient had remained entirely asymptomatic despite having an extremely severe form of Ebstein's anomaly which seems extremely unlikely considering the natural history of the disease and had maintained near normal saturation until now. The patient had been able to tolerate 3 full term vaginal deliveries successfully and given birth to healthy children in also a surprise especially if one considers the severity of her disease after having seen her echocardiogram and the severity of her anomaly. The other big surprise was that the patient had maintained normal saturation and left to right shunting across the ASD until this age. Various electrocardiographic patterns have been described in cases of Ebstein's anomaly but the pattern of an evolved inferior wall myocardial infarction pattern with no other conduction anomalies or evidence of pre-excitation has never been described previously

CONCLUSION

Rare presentations of Ebstein's late in adulthood form an important part of the spectrum and should form a part of differential diagnosis among clinicians and cardiologists dealing with patients presenting with similar symptoms in adulthood. This case highlights that Ebstein's can present as angina with an electrocardiogram which might corroborate the diagnosis and create a diagnostic dilemma. Early diagnosis and appropriate management

and surgical correction depending upon the severity of the lesion and the time of presentation form the key to the management of these cases. With better management strategies, it is hoped that survival of patients with this anomaly and the extent of cases picked up early in infancy will continue to improve.

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VIDEO LEGENDS

Video 1: apical four chambered view with color compare showing grossly dilated right atrium and right ventricle with compressed left ventricle and colorDoppler signal showing severe tricuspid regurgitation

Video 2: parasternal short axis view showing dilated right ventricle with the left ventricle compressed by the overloaded ventricle

Video 3: colorDoppler across the atrial septal defect showing the presence of left to right shunt

Video's are available in online only

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Angina and Pseudo infarct ECG Pattern in Ebstein's Anomaly

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