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Right Heart Failure Indicative of Ebstein's Disease in Adulthood about a Case

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Abstract

Ebstein disease is a rare congenital malformation whose clinical presentation differs according to the anatomical form and age of the patient. In adults, it presents mainly in the form of right or global heart failure or rhythm disorders. Survival is exceptionally long in some forms. We report the case of a 70-year-old patient with Ebstein's disease in right heart failure.

Keywords

Ebstein's Disease, Right Heart Failure

1. Introduction

Ebstein's malformation or abnormality (AE) is a congenital and complex heart disease of the tricuspid valve and right ventricle. It affects about 1/200,000 live births and occupies less than 1% of congenital heart disease [1]. It is characterized by apical displacement of the valvular ring, defect of delamination of the septal and posterior valves and often veil deformation of the anterior valve. This abnormal displacement divides the right ventricle into an atrial segment and a functional segment.

It was first described by Wilhelm Epstein in 1866 [2]. It is present at birth but symptoms can occur at any age, with an average life expectancy of the third decade, as indicated in an early report [3]. The clinical presentation varies according to the period of discovery, ranging from the neonatal form, very serious, to the better-tolerated forms of adolescents and adults [4]. In adulthood, it typically manifests as an arrhythmia or right or left heart failure [5]. Our goal is to

report a case of Epstein's anomaly in a 70-year-old woman, observed at the Chino-Guinenne Friendship Hospital in Conakry.

2. Observation

The case was a 70-year-old out-of-school patient living in a rural area admitted for bilateral oedema of both lower limbs, abdominal bloating, and exertional dyspnea. The examination showed spontaneous turgor of the jugular veins, sensitive hepatomegaly with hepatojugular reflux, distended abdomen, site of as-cites of medium abundance, blood pressure was 110/70mmHg, heart rate at 90 beats/min, Spo2 at 92% in ambient air; cardiac auscultation revealed cardiac arrhythmia with a mesocardiac systolic murmur, there was no cyanosis or digital Hippocratism; the lungs were free (Figure 1).

Front chest X-ray: Showed cardiomegaly with supradiaphragmatic tip and cardiothoracic index at 0.7 with hyperconvexity of the lower right arch. The pulmonary parenchyma was unremarkable (Figure 2).

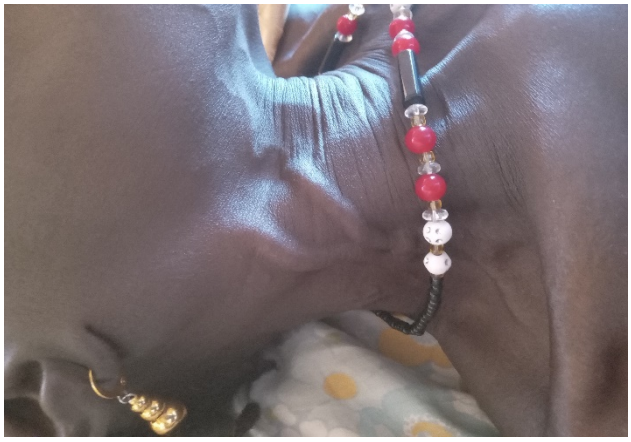

Figure 1. Spontaneous turgor of jugular veins.

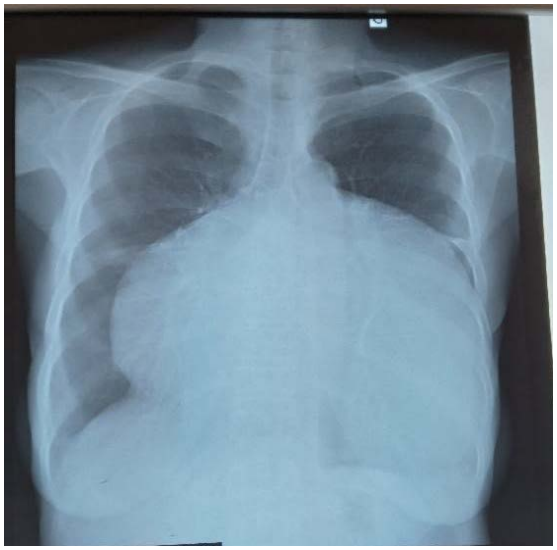

Figure 2. A frontal telethorax showing cardiomegaly with a significant right overhang.

The electrocardiogram:

Inscribed an irregular non-sinus rhythm with an average heart rate at 70 cycles per minute; Absent P-waves replaced by baseline tremulation concluding atrial fibrillation (Figure 3).

Echocardiography:

Reveals a low insertion of the septal and posterior leaflet of the tricuspid valve 15 mm below the tricuspid ring, a significant dilation of the right atrium by atrialization of the right ventricle (VD). The rudimentary VD and its atrial stump are dilated and responsible for compression of the VG (Figure 4).

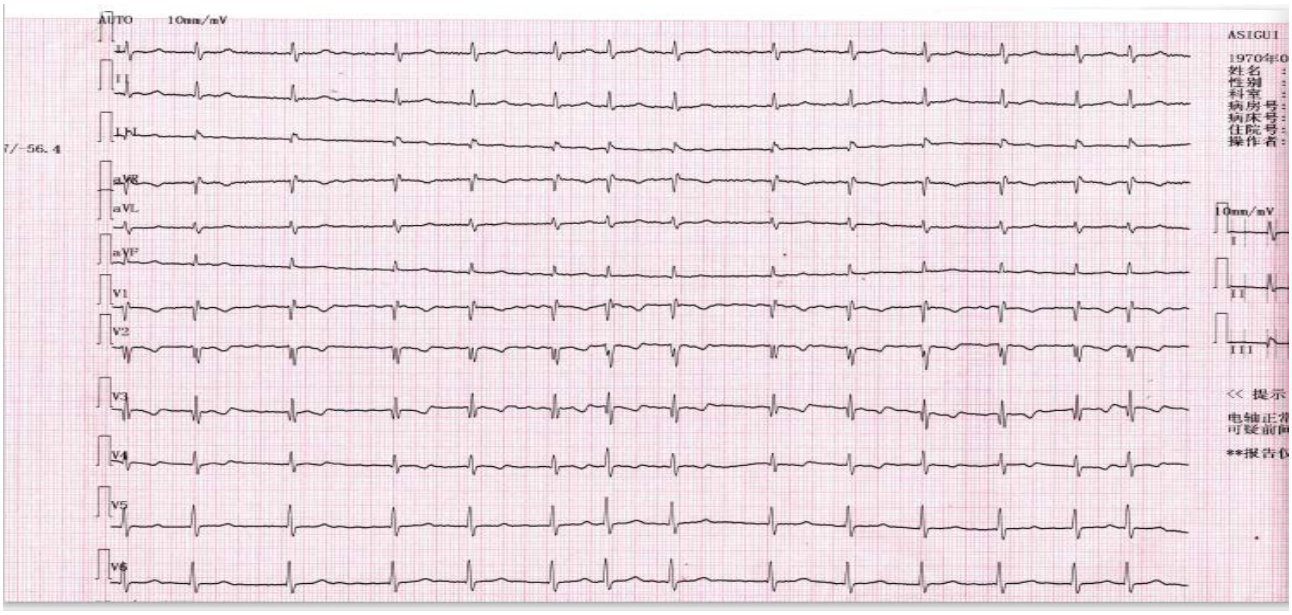

Figure 3. Electrocardiogram showing atrial fibrillation.

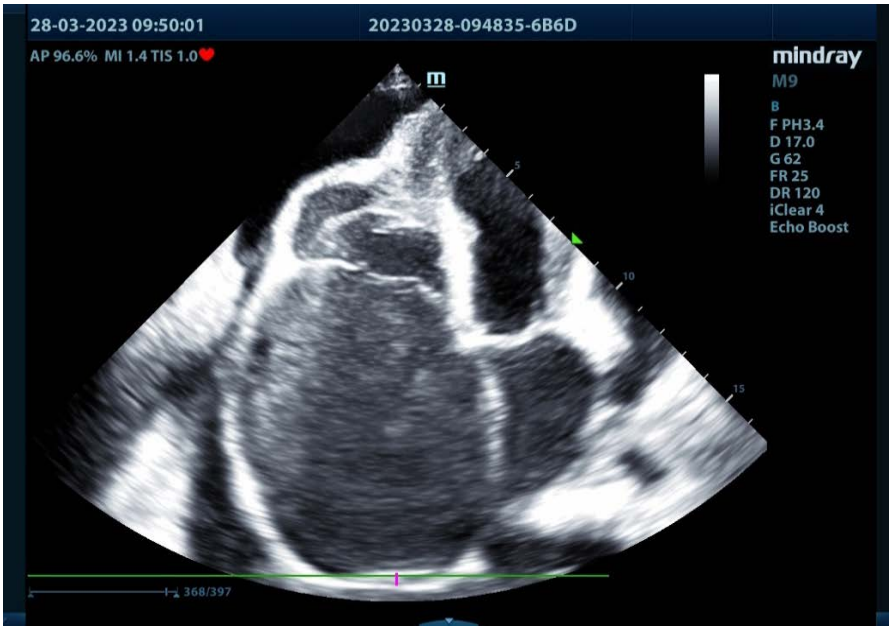

Figure 4. Echocardiography of Ebstein's anomaly.

3. Discussion

Ebstein's disease is a congenital anomaly of the tricuspid valve whose septal and posterior leaflets are attached to the ventricular wall and moved to the tip of the right ventricle [6]. These anatomical abnormalities divide the right ventricle into a thin-walled proximal portion that atrial and widens, and a more distal trabeculate component that forms the functional right ventricle [7]. The pathophysiological consequences of this malformation are tricuspid insufficiency, intra-atrial mechanical desynchronization by sequential activation of the true atrial segment and the atrial ventricular segment and therefore responsible for right heart failure as illustrated by our patient; The clinical presentation is variable and depends on age and anatomical form, usually diagnosed on the occasion of cyanosis, right heart failure or cardiac arrhythmia.

Heart rhythm disorders are the prerogative of elderly patients [8], as reported in our clinical case where the patient was in atrial fibrillation, due according to some authors to the dilation of the right atrium and fibrous degerescence of the myocardium [9] [10]; in the study by Antit *et al.* in Tunisia in 2021 on the electrocardiographic aspects of Ebstein's disease, atrial fibrillation accounted for 7% of cases.

Transthoracic echocardiography is the paraclinical examination determining in the diagnosis of AE, it allows an accurate evaluation of the anatomy of the tricuspid valve, the size and contractility of the VD, the search for other associated heart defects. It is also used to determine prognosis through the Celermajer index. Our case was an isolated form.

Management of AE depends on the patient's age, severity of anatomical damage, associated heart defects and clinical presentation. For centers with adequate technical platforms, patients who have a type of Celermajer ≥ B may benefit from surgical repair [11] [1]. Our patient did not receive surgical treatment; Treatment of right heart failure involves adjusting heart rate and preload and restricting exercise. This may include a low-sodium diet, oral diuretics, digoxin and a low-dose angiotensin-converting enzyme (ACE) inhibitor. The management of the right heart failure flare-up is mainly based on diuretics, vasodilators, cardiotonics, low-sodium diet [12]. Our patient was treated with diuretics, angiotensin-converting enzyme inhibitor and anticoagulant.

The natural course of the disease depends on the degree of displacement and attachment of the tricuspid valve and myocardial involvement causing two main clinical presentations, the severe perinatal form, and the more or moderate form of children and adults [13]. In our patient, the evolution was favorable with a good regression of the congestive signs of right heart failure and a slowdown in ventricular cadence followed by hemodynamic improvement.

4. Conclusion

Ebstein's anomaly is a complex congenital heart disease with several clinical presentations. The forms age are not. Symptomatic medical treatment of Ebs-

tein's malformation allows in certain circumstances to improve the prognosis of the disease.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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