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An Uncommon Association of Ebstein's Anomaly and Acute Rhumatic Fever

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Abstract **Case Report**

Background: Ebstein's anomaly is a relatively rare congenital heart disease (atrialisation of the right ventricle). Anomalies of left side of the heart have been described with this condition. We however present a case of Ebstein's anomaly with recurrent acute rheumatic fever (ARF) and mitral stenosis. ***Case report:*** A 13-year-old boy with not followed for heart disease and with a history of notion of repetitive angina, is admitted to the pediatric emergency room for fever and polyarthritis. General examination not showed subcutaneous nodules and laboratory investigations revealed leucocytosis, elevated erythrocyte sedimentation rate (ESR) and antistreptolysin O (ASO) titre. Echocardiography revealed Ebstein anomaly with moderate mitral stenosis. A diagnosis of recurrent ARF was made based on the carditis, arthritis, history of recurrence of sore throat and evidence of recent streptococcal infection. He was transferred to the cardiology department and initially put on medical treatment. Its clinical course was marked by the appearance of unresolved ventricular tachycardia despite drug cardioversion. His prognosis was unfavorable with death after 10 days of hospitalization under antiarrhythmic treatment. ***Conclusion***: Ebstein's disease is a rare disease. Cardiac arrhythmias are deleterious and the association of rheumatic fever with co-morbidity increases its lethality.

Keywords: Ebsteins anamoly, Rheumatic mitral stenosis, Tricuspid regurgitation.

INTRODUCTION

Ebstein's anomaly (EA) is a rare congenital malformation of the tricuspid valve (TV) with myopathy of the right ventricle (RV) that has variable presentation of the anatomic and pathophysiologic characteristics [1]. The EA accounts for <1% of all congenital heart defects (CHDs), with a prevalence of 1 per 200,000 live births [2].

This malformation includes electrical conduction abnormalities and supraventricular arrhythmias. Heart disease itself can be complicated by rheumatic heart disease, especially in underdeveloped countries with a high incidence of strep infection.

Acute rheumatic fever (RF) is a real public health problem in Africa, with 13.3% of the population in the developing world may be a carrier of RF from the throat, and nearly 40% of those infected may suffer from the tragic complication, rheumatic heart disease (RHD). The incidence of RHD in Africa is estimated at 17-43% of all cardiovascular diseases [3]. In Morocco the incidence of rheumatic carditis has been estimated at 53.1% [4].

However Rheumatic heart disease associated with Ebstein's anomaly of the tricuspid valve is a very rare clinical entity with only a handful number of cases reported in literature. Here we report an association of rheumatic fever with Ebstein's disease associated with mitral rheumatic heart diseases which was complicated by cardiac arrhythmias fatal to the patient.

CASE REPORT

A 13-year-old boy, he presented as antecedent a notion of repetitive angina with a last episode dating back one month.

He was first admitted to the pediatric emergency room for polyarthralgia, temporary, migratory, painful knees and ankles. He also reported a notion of fever, dyspnea on exertion classified stage III by the NYHA and deterioration of general condition.

The clinical examination on admission revealed a conscious stable , feverish patient with a temperature of 39 ° Celsius, an arterial pressure at 110/70 mmhg, a tachycardia at 100 beats/min , a saturation at 91% in the open air , a respiratory rate at 23 cycles/min. The lung exam is normal. Cardiovascular examination revealed a systolic murmur at the tricuspid focal point with a nominal intensity of 4/6 and a diastolic bearing at the mitral focal point.

Physical examination finds signs of right heart failure: turgor of jugular veins, hepato-jugular reflux, hepatomegaly, positive Harzer's sign, and retro-malleolar edema of the lower limbs white, soft.

The laboratory workup revealed a biological inflammatory syndrome with white blood cells at 28000 /uL, C-reactive protein at 230 mg / l, sedimentation rate at 96 mm / hour , hepatic cytolysis ASAT at 248 U / L and ALAT at 1154 U / L (hepatic shock) with ASLO at 1170 U / L (Table 1).

The twelve-lead EKG (Figure 1) showed sinus tachycardia at 110 beats/ min with an enlarged right atrium and incomplete right bundle branch block. A chest x-ray (Figure 2) showed a cardiomegaly with a rectilinear left middle arch.

Plus, the transthoracic echocardiography (Figure 3) showed an apical insertion of the septal valve with a defect in valvular coaptation and ring dilation resulting in tricuspid laminar insufficiency, and moderate mitral stenosis. The dilated right ventricle has good systolic function. The left ventricle of normal size and systolic function preserved. All associated with a blade of pericardial effusion. This echocardiographic appearance is characteristic of Ebstein's disease.

In addition, the diagnosis of rheumatic carditis posed: history of recurrent angina with a last episode dating back one month, migratory polyarthritis of the large joints, fever, positive laboratory workup, positive ASLO associated with moderate mitral stenosis and pericardial effusion. The child was transferred to the cardiology department for further care.

Initially put on injectable lasilix 40 mg per day, aldactone 25 mg, cortancyl 60 mg, and potassium syrup, calcined and antibiotic therapy.

The surgical indication for a tricuspid plasty was made after stabilization of the patient.

On the fifth day of his hospitalization, he presented with abrupt onset and end of resting palpitations, well tolerated with arterial pressure of 115 / 70mmhg without signs of hemodynamic instability.

A twelve-lead electrocardiogram (Figure 4) showed a regular tachycardia with wide QRS at 170 bpm, duration of QRS complex at 160 ms, RS duration at 160 ms in V2, initial R wave in AVR, Vi / Vt <1, the diagnosis of ventricular tachycardia posed.

We initiated a loading dose of cordarone followed by a maintenance dose. After a return to sinus rhythm, we observed short PR interval at 60ms, an initial empatment of the QRS which is the delta wave.

The accessory bundle has been diagnosed: short PR interval at 60ms, an initial empatment of the QRS which is the delta wave (Figure 5). Removal of the accessory bundle in a specialized center was asked.

The clinical and biological evolution was satisfactory with apyrexia, disappearance of congestive signs and regression of the inflammatory balance with white blood cells at 15540 /uL, the Creative protein at 44 mg / l as well as the ASAT liver test at 32 U / L and ALAT at 22 U / L.

On the twelfth day of his hospitalization, the patient died of hemodynamic instability associated with palpitations at rest, shocked repeatedly without success.

Table 1: Result of laboratory test

Complete blood count		**Biochemistry**	
Hemoglobin	14.7 g/dL	Na	137mmol/L
Hematocrite	43 %	K	4,2 mmol/L
Leucocyte count	28×10^3/uL	Cl	94 mmol/L
Platelet count	543×10^3/uL	Creatinine	3,9 mg/L
Reticulocyte	11730 /uL	Urée	0,22 g/L
Neutrophils	23.58×10^9/L	ALT	248 U/L
Lymphocytes	3.03×10^9/L	AST	1154 U/L
Monocytes	1.57×10^9/L	Ca2+	94 mg/L
Eosinophils	0.01×10^9/L	Phosphore	41 U/L
		Vitamine D	3.08 ng/L
Inflammatory markers		ASLO	1470 U/ML
VS	96 mm/h	Albumine	38 g/L
CRP	230 mg/L	TSH	0,58 mUI/L
		T3	3.1 pg/mL

Complete blood count		**Biochemistry**	
Serologie		T4	17,3 pg/mL
HIV antibody	Negative	LDH	48 6 UI/L
Hepatic serology	Negatif	Bicarbontes	14.8 mmol/L
syphilitic serology	Negatif	Glycémie	1.1g/l

CRP: C-reactive Protein; VS: Erythrocyte Sedimentation Rate; HI: Human Immunodeficiency

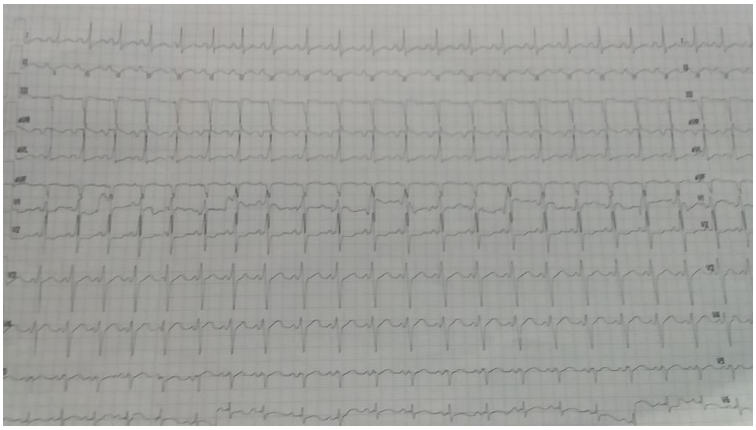

Figure 1: Showed sinus tachycardia at 110 beats /min with an enlarged right atrium and incomplete right bundle branch block

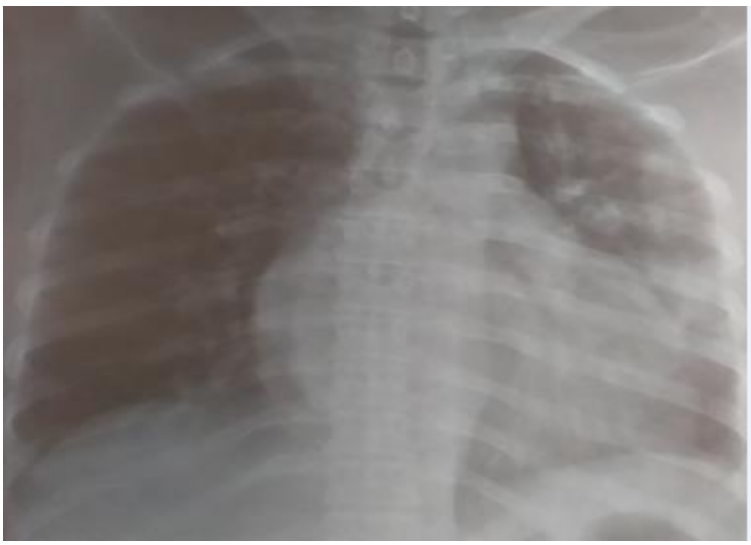

Figure 2: Chest x-ray found cardiomegaly with a rectilinear left middle arch

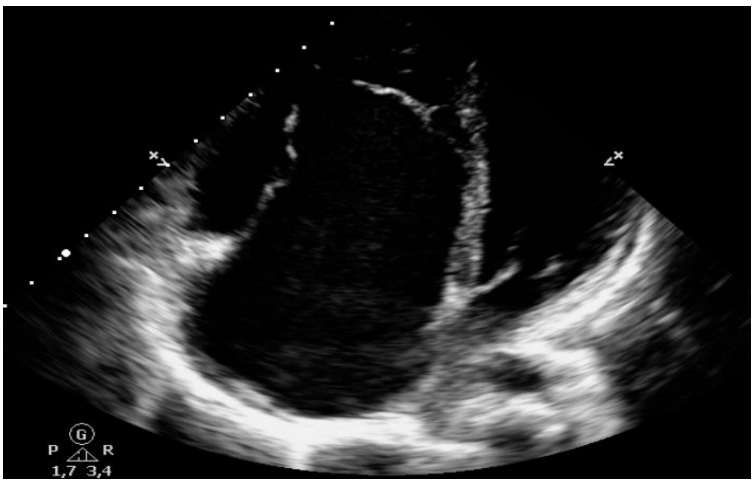

Figure 3: Showed an apical insertion of the septal valve with a defect in valvular coaptation and ring dilation resulting in tricuspid laminar insufficiency, and moderate mitral stenosis

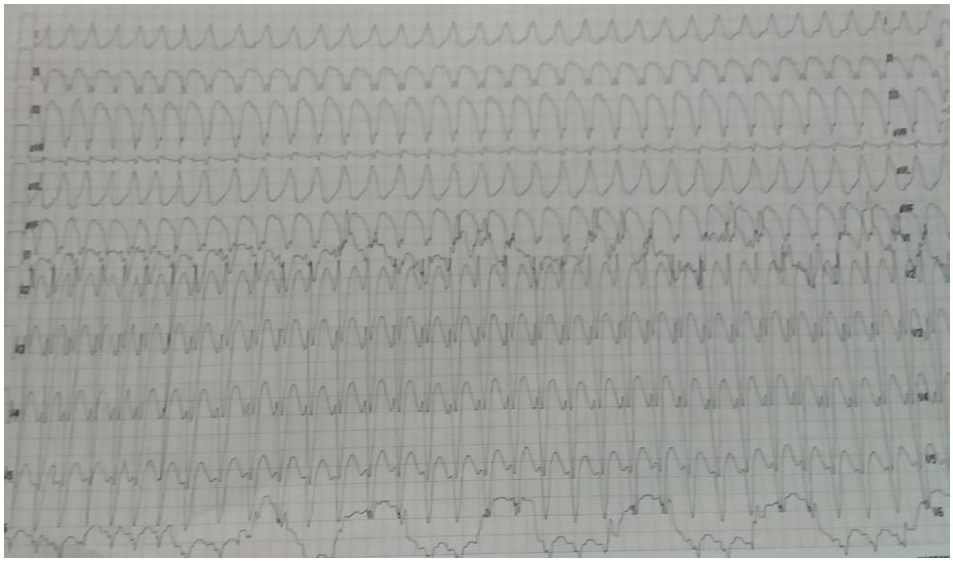

Figure 4: A regular tachycardia with wide QRS at 170 bpm,duration of QRS complex at 160 ms, RS duration at 160 ms in V2, initial R wave in AVR, Vi / Vt <1 : the diagnosis of ventricular tachycardia posed

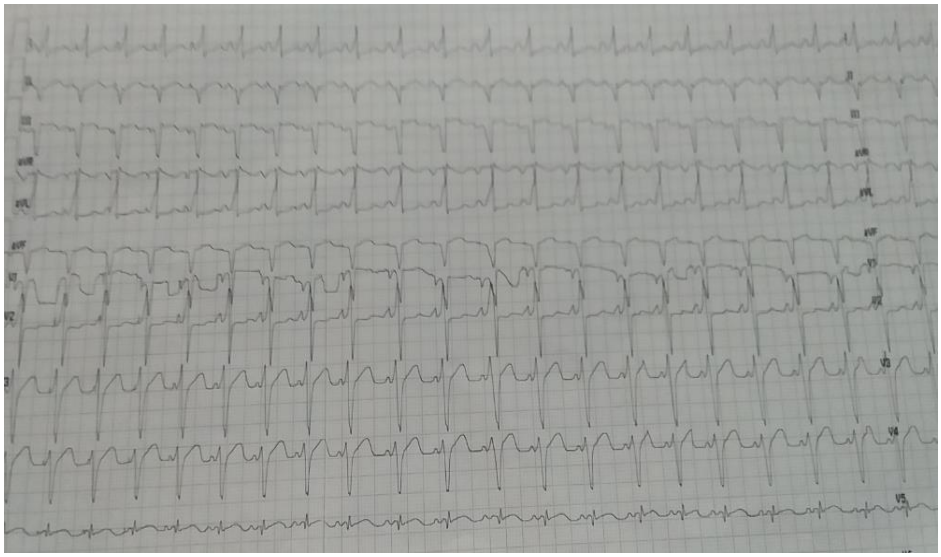

Figure 5: Accessory bundle has been diagnosed: short PR interval at 60ms, an initial empatment of the QRS which is the delta wave

DISCUSSION

Ebstein's anomaly (EA) is a rare congenital heart disorder affecting tricuspid valve causing regurgitation with an incidence rate of 1 per 2,00,000 live births and accounting for 1% of all cases of congenital heart disease [5].

EA is a malformation of the tricuspid valve and right ventricle, which encompasses a large spectrum of disease severity. There is failure of delamination of the septal and posterior tricuspid valve leaflets from the myocardium, resulting in apical displacement of the tricuspid valve ($\geq$0.8 cm/m^2 relative to the anterior mitral leaflet insertion site). The portion of the right ventricle proximal to the functional tricuspid valve becomes "atrialized." The anterior leaflet of the tricuspid valve is large and redundant, and often has fenestrations and tethering attachments to the right ventricular (RV) free wall. The posterior leaflet is often dysplastic and atrially displaced [6].

The clinical presentation of Ebstein anomaly varies widely. In general, symptoms are related to the degree of anatomic abnormality.

In a review of 220 patients with Ebstein anomaly, early age at presentation was frequently associated with other cardiac lesions, particularly atrial septal defect and pulmonary stenosis, which predisposes to cyanosis from right-to-left shunting [7].

In children, adolescents and adults , symptoms such as exertional dyspnea, fatigue, cyanosis, and palpitation may be occur; these symptom may be due to right ventricular dysfonction, severe tricuspid valve regurgitation or right-to-left shunt at the atrial level. Atrial tachyarrhythmias are present in approximately 20

to 30 percent of cases across age groups, with greater frequency in adolescents and adults [7, 8]. Some of these arrhythmias may be due to accessory conduction pathway(s), present in up to 20 percent of patients; the majority of these pathways are located around the orifice of the malformed tricuspid valve [9-11].

An ECG is not required for the diagnosis of Ebstein anomaly but is an important component of the overall evaluation of patients with this condition to detect arrhythmias and evidence of preexcitation [12].

The chest radiograph results vary depending upon the severity of the disease [13]. In severe cases, reveals massive cardiomegaly (often termed a "wall-to-wall" heart) with a small ascending aorta and normal or diminished pulmonary vascularity like our patient.

Comprehensive transthoracic two-dimensional and Doppler echocardiography is the most useful tool for establishing a diagnosis of Ebstein anomaly. The key diagnostic finding for Ebstein anomaly is the apical displacement of the septal tricuspid valve leaflet indexed to the body surface area (by ≥8 mm/m [compared with the position of the anterior mitral valve leaflet]) demonstrated in the apical four-chamber view. The degree of displacement affects the severity of clinical manifestations [14, 15].

Cardiovascular magnetic resonance is complementary to that obtained from echocardiography, combined CMR and echocardiographic imaging is usually performed prior to and following surgical tricuspid valve intervention [16, 17].

The anatomical variation of the tricuspid valve in Ebsteins anamoly increases the risk of accessory atrioventricular connections and pre-excitation. 6%–36% of these patients have accessory pathways. Syncope and sudden death have been reported and may be caused by atrial fibrillation with a rapid ventricular response due to fast conduction through an accessory pathway or from ventricular arrhythmias. Arrhythmias may also occasionally precipitate heart failure [1].

The components of management are monitoring, medical management (including temporizing relief of symptoms prior to surgery), management of arrhythmias, and surgical or catheter intervention.

Rheumatic heart disease (RHD) affects 33 million people worldwide and results in approximately 320,000 deaths annually. It remains a crucial concern of cardiovascular death, particularly in countries with less resources, where RHD frequently causes high morbidity and mortality [18, 19].

Rheumatic heart disease is the most serious sequela of rheumatic fever, developing to a varying degree in up to 50% of patients with rheumatic fever, leading to mitral valve stenosis early or later in life. It may take several years after an episode of rheumatic fever for valve damage to develop or symptoms to appear [20].

Rheumatic heart disease associated with ebstein's diseases is an extremely rare clinical entity.

In our reported case, our patient had associated moderate mitral stenosis. Mitral valve abnormalities have been described in Ebstein's anomaly, but acquired rheumatic mitral valve disease is an extremely rare association;

CONCLUSION

Ebstein's disease is a rare entity. Rheumatic disease remains a real development problem in our countries.

Their combination is rare and can lead to the early onset of unwanted complications, including increased rhythmic risk, pulmonary hypertension and heart failure.

Conflict of Interests: There are no conflicts of interests for the development of this publication.

Ethical Standards: Informed consent was obtained from the patient's parents for the publication of this case

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