Case Report

Adult Ebstein Anomaly with Right Atrial Myxoma

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ABSTRACT:

BACKGROUND:

Ebstein Anomaly is rare in adults; a combination of this disease with cardiac myxoma has not been reported previously.

METHODS:

Surgery was performed by removal of the myxoma and replacement of the tricuspid valve and correction of the abnormal atrialization of the right ventricle.

RESULTS:

The patient had a smooth postoperative recovery and improved dramatically after wards.

CONCLUSION:

We have dealt with this rare condition successfully and the patient had good outcome.

KEY WORDS: Adult Ebstein Anomaly, Cardiac myxoma with Ebstein Anomaly, surgery on Ebstein Anomaly.

INTRODUCTION:

In 1866, Dr. Wilhelm Ebstein, a young physician in Breslau, Poland, described the unusual cardiac findings in a 19-year old laborer, who had died of cyanotic heart disease. The anterior leaflet of the tricuspid valve was enlarged and fenestrated and there was downward displacement of the posterior and septal leaflets in a spiral manner below the annulus, they were hypoplastic, thickened and adherent to the wall of the right ventricle [1].

Ebstein Anomaly (EA) is a rare cardiac anomaly that accounts for less than 1% of all congenital heart diseases; it involves both sexes equally [1]. *Although few patients reach advance age, life expectancy for most is limited*, and the most common cause of death is congestive heart failure, hypoxia and cardiac arrhythmia [1, 2, 3, 4, 5].

When diagnosis is made in infancy, the prognosis is worse; one-third to one-half of these patients will die before 2 years of age [4].

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Ebstein Anomaly has three characteristics [1, 2, 4]:

1. Downward displacement of septal and posterior leaflets into right ventricle and sail-like abnormality of the anterior leaflet, this displacement leaves a portion of the ventricle above the valve as an integrated part of the right atrium (atrialized ventricle).
2. An atrialized portion of the right ventricle between the tricuspid annulus and the attachment of the posterior and septal leaflets.
3. A mal formation of the right ventricle with a marked reduction in its size.

So according to Carpentier, it is classified into 3 types [4]:

A. Valve insufficiency with normal leaflet motion.

B. Valve insufficiency with leaflet prolapse.

C. Valve insufficiency with restricted leaflet motion.

Although important stenosis is uncommon, most Ebstein valves are incompetent, often severely so. This is contributed to by marked dilatation of the true tricuspid annulus and the right ventricle, as well as the morphologic abnormalities of the tricuspid valve [3].

The right atrium is enormously dilated in advance cases and usually an interatrial communication (cause paroxysmal embolization in adult) present in most cases as Patent Foramen Ovale (PFO), although ASD may be present and rarely ostium primum coexist [2, 3,].

Symptoms are related to severity of the incompetence of the tricuspid valve, presence or absence of associated Atrial Septal Defect (ASD), impairment of right ventricular function and presence of associated cardiac anomaly, like right cor-triatriatum and right overt accessory atrioventricular pathway [1, 2, 6].

In older patients, the predominant symptoms are fatigability, dyspnea on exertion and cyanosis. Palpitation in the form of paroxysmal atrial arrhythmia and premature ventricular beats are common, less frequently ascitis and peripheral edema are present [1, 2, 3, 7, 8].

Physical signs vary, heart sounds are usually soft, and a multiplicity of sounds and murmur is often

heard, all originating from the right side of the heart, a systolic murmur of tricuspid regurgitation may be heard along the left sternal border [1, 2, 3, 4]. Low intensity diastolic and presystolic murmur result from anatomic or functional tricuspid stenosis [1, 2, 3, 4].

Diagnosis of Ebstein Anomaly; usually follows the routine steps in diagnosing cardiac diseases, starting with the essential chest x-ray and ECG, to show the marked atrial dilatation and cardiomegally [1, 2, 3, 4]. Echocardiography, is essential to study the tricuspid valve and to show the displacement of the septal and posterior leaflets and to check the atrialized right ventricle and its size and the left ventricular function with the advanced use of the Live/real time three-dimensional transthoracic echo [9, 10].

Cardiac catheterization is reserved only for cases which seem difficult to assess by echo study alone, so we can study well the tricuspid valve and the severity of its incompetence and to check the coronaries for any associated lesions, especially in adult cases [1, 2, 3, 4].

CASE STUDY:

Male patient (A. J.) 45 years old referred to the surgical dept. in Ibn Al-Bitar Hospital for Cardiac Surgery on March 2005, with hemoptysis and features of severe right sided failure including; lower leg edema, ascitis, palpable liver and pleural effusion which necessitated frequent pleural aspirations.

On reception, routine investigations were done for him like chest x-ray & ct scan of the chest (fig. 1); showed a huge heart and an ECG which showed Atrial Fibrillation (AF).

Abdominal and chest Ultrasonography (fig. 2) showed hepatosplenomegaly, ascitis, left pleural effusion, engorged Inferior Vena Cava (IVC) and thickened Gall Bladder. Transthoracic Echocardiography -TTE (fig. 3) study showed the following data; Aortic root dimension 25mm, Left Ventricular Diastolic Dimension (LVDD) 45mm, Left Atrial dimension (LA) 33mm, Left Ventricular Systolic Dimension (LVSD) 30mm and Ejection Fraction of 62%.

Transesophageal Echocardiography (TEE) study labeled the patient as tricuspid valve regurgitation (Ebstein Anomaly) with sail-like anterior leaflet adherent to the wall, rudimentary posterior and septal leaflets with downward displacement of 28mm from the annulus. Cardiac catheterization (fig. 4), confirmed Ebstein Anomaly.

On May 2006, surgery was performed which showed two small atrial masses near the opening of the Superior Vena Cava (SVC) of about 1 x 0.5 cm and a very dilated and incompetent tricuspid valve (type C according to Carpentier).

A tissue valve (BICORE ^{TM} – SJM size 33) was inserted according to the method described previously[11] which entitle utilizing the anterior leaflet of the tricuspid valve to attach the biological valve anteriorly and the opposite area of the atrialized ventricle is used for the attachment of the rest of the valve, and the rest of apparatus of the tricuspid valve was left without interference. The patient ran a smooth postoperative period apart from hypoproteinemia and high fluid demand which was corrected by intravenous albumin, plasma and colloids.

Early follow up after a week revealed marked improvement in the general condition with decrease in the edema and sound decrement in cardiac size by CXR and ECG (fig. 5), TTE (fig. 6) showed well functioning competent tissue valve and rebuilding of the atrialized part of the right ventricle and improvement in right ventricular contraction with good left ventricular function.

Later on, histopathology of the mass reported the presence of myxoma as a benign tumor in the right atrium (fig. 7).

DISCUSSION & CONCLUSION:

Ebstein Anomaly is a rare congenital heart disease and can be manifested in adults as well as in children [1, 2, 3, 4, 11]. Diagnosis of EA is usually a straight forward procedure and can follow the usual diagnostic steps of all cardiac diseases [1, 2, 3, 4, 9, 10].

Association of EA with other cardiac diseases is reported [1, 2, 3, 4,], and it may add to the severe symptoms of right sided failure and congestive heart failure, and in our

Patient, there were *a unique association of EA with Right Atrial Myxoma*, in which as far as our knowledge, is reported for the first time, as there was a report of an EA with cardiac rhabdomyomata [12].

Surgery for EA is recommended by many authors, and it ranges from repair of the tricuspid valve to total replacement or to new methods of inserting valves [1, 2, 3, 4, 11, 13, 14, 15].

Usually the results are good, and the patients seems to benefit well from the surgery in all types, apart from the expected possible complications of right sided failure which may take longer time to resolve on heavy diuretic therapy and cardiac inotropic support, also regarding right sided prosthetic valves thrombotic complications due to low pressure circulation [1, 2, 3, 4, 11, 13, 14, 15].

So in conclusion; we had dealt with unusual adult EA associated with atrial myxoma which was not diagnosed preoperatively, surgery was a success.

Legends:

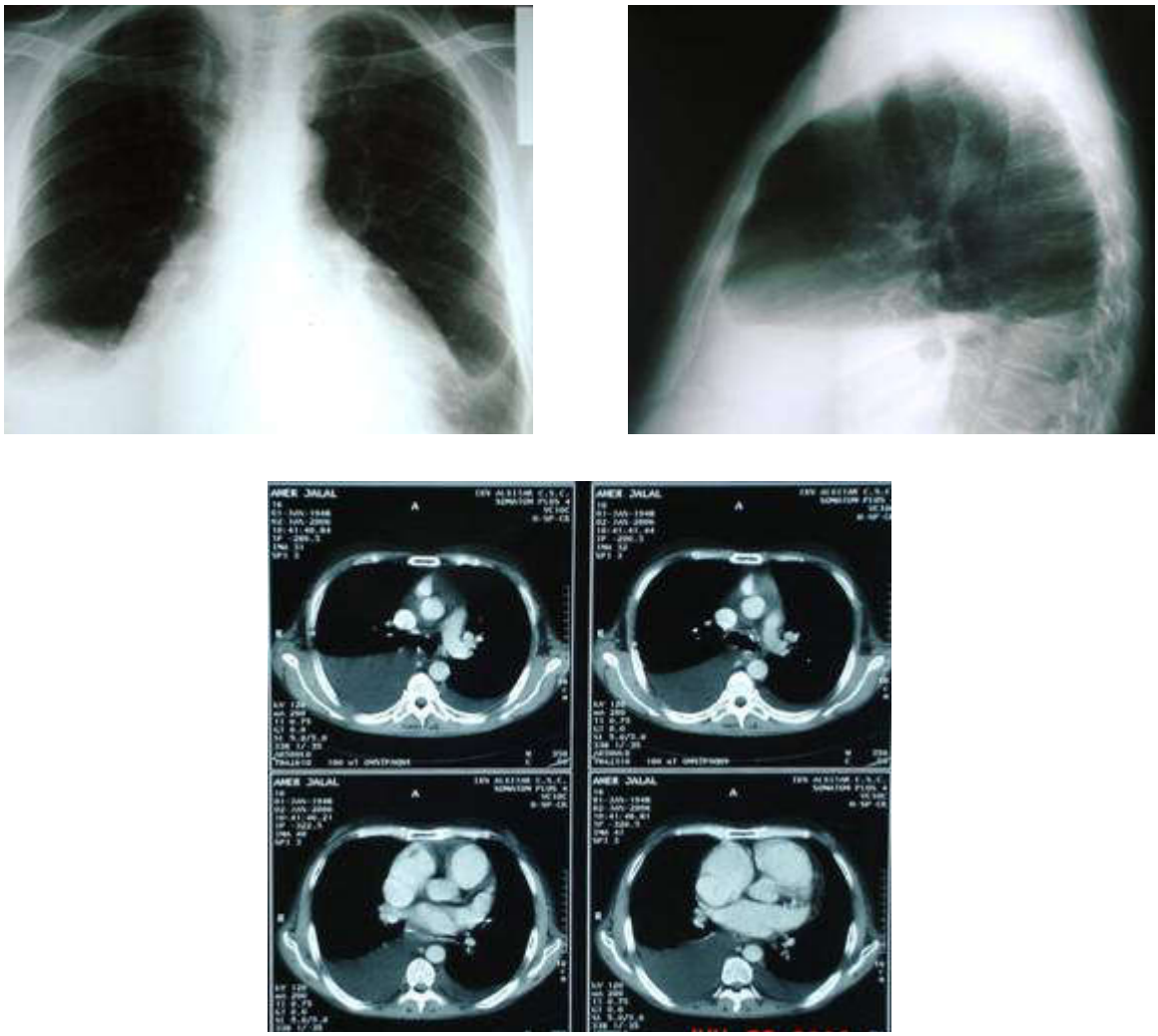

Fig. 1, Preoperative chest x-rays and ct-scan showing huge heart with with evidence of pleural effusion

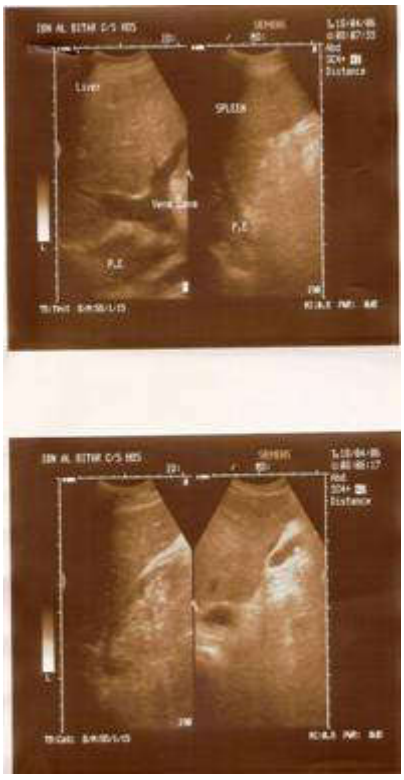

Fig. 2, Abdominal US showing engorged IVC and hepatomegaly

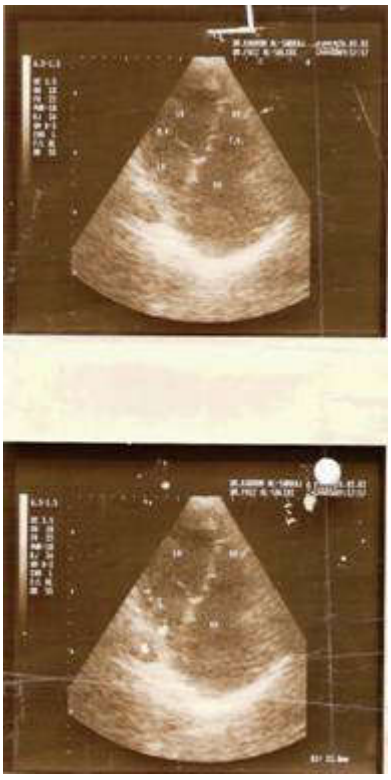

Fig. 3, Pre operative TTE

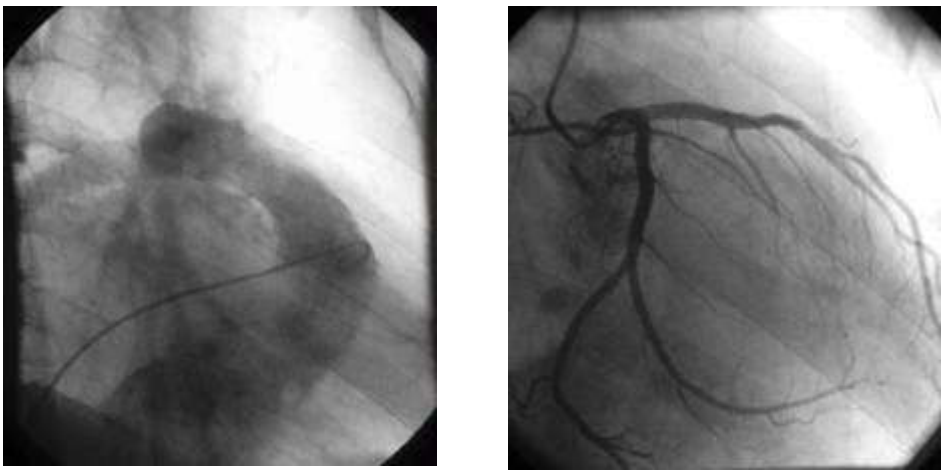

Fig. 4, Preoperative cardiac catheterization

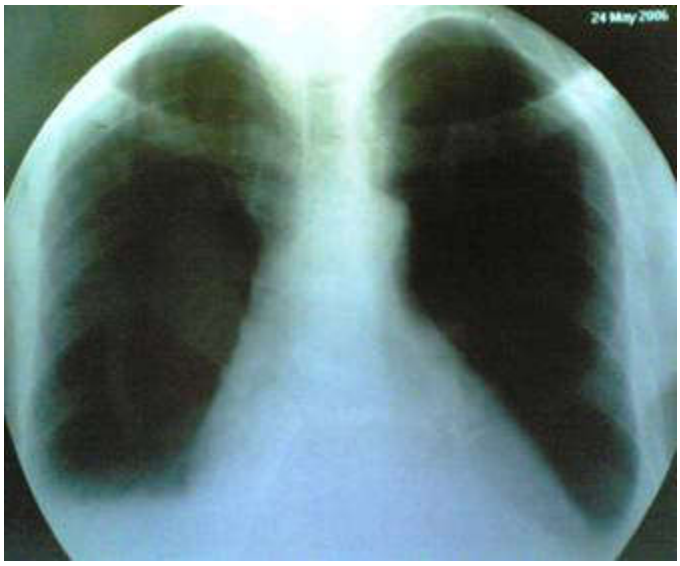

Fig. 5, Early Postoperative Chest X-ray

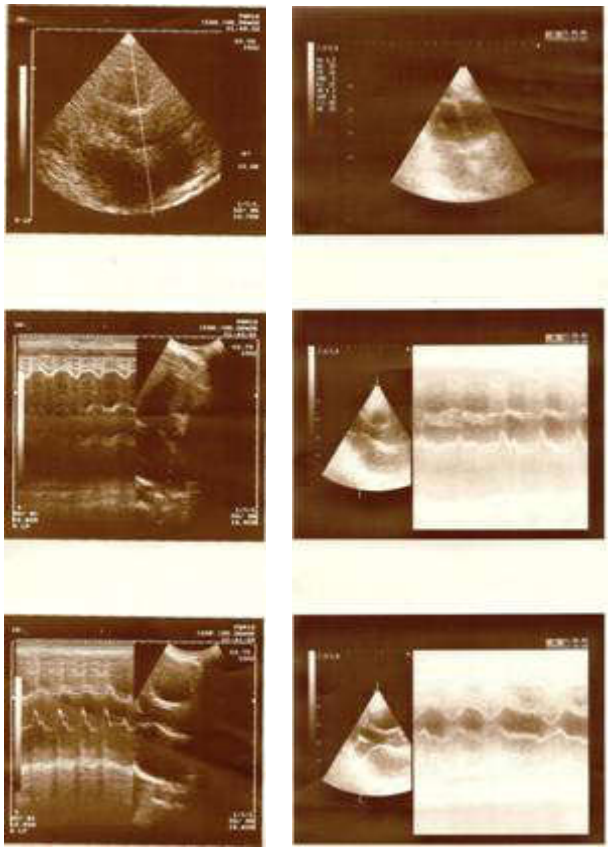

Fig. 6, Postoperative ECHO

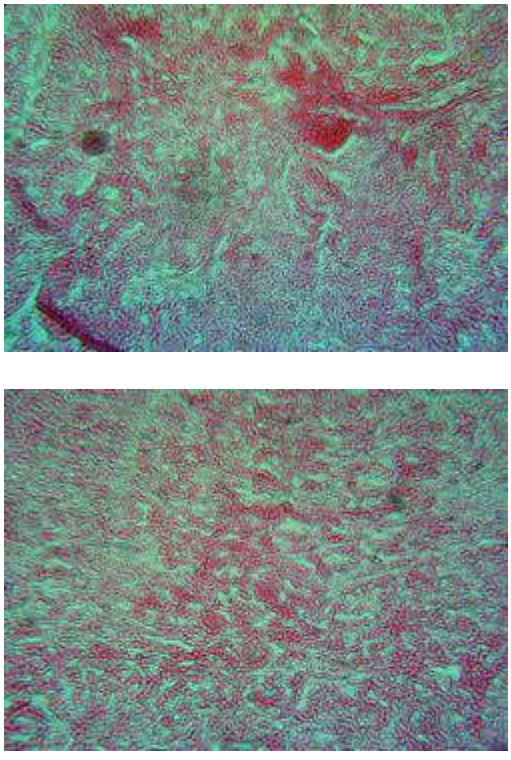

Fig. 7, Histopathological result of the right atrial mass showing the classical picture of myxoma

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