Ebstein's Anomaly with Non-Patent Ductus Arteriosus Aneurysm and Bradycardiac Atrial Fibrillation in an 80-year-old Man

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An 80-year-old man with Ebstein's anomaly and ductus arteriosus aneurysm is reported. He was admitted with bradycardiac atrial fibrillation and right ventricular failure. For the control of brady-arrhythmia, a permanent pacemaker was implanted. Two-dimensional echocardiogram revealed distal displacement of the septal tricuspid valve. Aortography and computed tomography showed ductus arteriosus aneurysm. This is the first report of the association of Ebstein's anomaly and non-patent ductus arteriosus aneurysm.

Key words: Pacemaker, Right ventricular failure, Echocardiogram

It is well known that the overall prognosis of Ebstein's anomaly is very poor and the survival rate is very low (1–3); only a few patients surviving for 70 yr or longer have been previously reported (4–6). Watson reported that of 505 cases of Ebstein's anomaly, 77 (13.3%) died from natural causes, and 31 (54.4%) of the 57 who had surgical treatment did not survive the operation (1). Ductus arteriosus aneurysm is a rare lesion, especially in adults (7–14). Of the approximately 70 cases of ductus arteriosus aneurysm in the literature, only 17 cases were in adult patients (7–14). We describe a rare case of the association of Ebstein's anomaly and ductus arteriosus aneurysm.

CASE REPORT

An 80-year-old man was admitted to Tottori University Hospital on October 6, 1988, complaining of back pain, oliguria and dyspnea. He had been in excellent health all of his life and had led a relatively illness-free life until 1978 (age 70), when he was first admitted to a hospital for acute myocardial infarction. From this time, he received diuretic therapy for the control of cardiomegaly. There was no history of chest trauma or sepsis.

Physical examination revealed an irregular pulse with a rate of 40 beats/min and blood pressure of 90/60 mmHg. The conjunctiva bulbi were slightly icteric, and there was a pronounced jugular vein pulsation with giant v waves and a normal y descent. On examination of the chest, cardiomegaly was observed, and the third heart sound was heard. There was no heart murmur or rales. Severe tricuspid regurgitation was suspected clinically. The liver was palpable 3 cm below the costal arch in the right midclavicular line, and was blunt and firm. There was no pretibial pitting edema.

Results of urinalysis, complete blood count and serologic test for syphilis were either within the normal limits or negative. Blood chemistry showed a creatinine level of 3.5 mg/dl and total bilirubin of 2.7 mg/dl. The electrocardiogram showed
bradycardiac atrial fibrillation (HR = 36/min) and complete right bundle branch block (Fig. 1). There were abnormal Q waves in II, III and aVF (Fig. 1). Chest X-ray disclosed cardiomegaly (cardio-thoracic ratio = 75%) with a somewhat globular appearance to the overall cardiac silhouette, and a smooth rounded density in the left suprahilar region contiguous with the aorta, which in the lateral view was projected on the aortopulmonary window (Fig. 2). Comparison with a chest film taken 4 yr prior to admission demonstrated the mass, but it was considerably smaller.

Apical four-chamber echocardiogram revealed that the septal tricuspid leaflet was markedly displaced downward into the right ventricle by 35 mm and thus the right ventricle was divided into two components, an atrialized right ventricle and a func-

Fig. 1. Electrocardiogram showing bradycardiac atrial fibrillation and complete right bundle branch block.

Fig. 2. Anteroposterior (left) and lateral (right) chest X-ray films show the left paraaortic mass in the superior mediastinum.

Fig. 3. Two-dimensional echocardiogram demonstrating apical four-chamber view from apex to base (top to bottom). Septal leaflet of tricuspid valve arises well below its annulus and is displaced into body of the right ventricle (35 mm displacement). AML, anterior mitral leaflet; ATL, anterior tricuspid leaflet; LA, left atrium; RA, right atrium; RV, right ventricle; STL, septal tricuspid leaflet.
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tional right ventricle (Fig. 3). Apical long-axis echocardiogram showed severe hypokinesis limited to the diaphragmatic wall of the left ventricle. Two-dimensional color Doppler echocardiogram showed the tricuspid regurgitant flow signal directed from the functional right ventricle into the atrialized right ventricle and the right atrium. No other cardiac malformation was observed. The diagnosis of Ebstein's anomaly and tricuspid regurgitation was made. Right cardiac catheterization disclosed the following pressures in mmHg: pulmonary artery, 22/4 (mean 11); right ventricle, 23/5; right atrium, 23/3 (ventricularization of pressure curve); superior vena cava, mean 15; inferior vena cava, mean 13. There were not any significant oxygen step-ups of oxygen saturation in the right atrium, the right ventricle, or the pulmonary artery. The presence of a left-to-right shunting heart disease was ruled out.

For the control of bradycardiac atrial fibrillation, temporary right ventricular pacing was started. From the second hospital day, the total daily urine volume increased. On the sixth hospital day, serum creatinine became within the normal limit (1.4 mg/dl), but hypotension and hepatomegaly continued. On the ninth hospital day, a permanent pacemaker was implanted. The electrocardiogram showed normal right ventricular inhibited (VVI) pacing (Fig. 4).

After surgery, the abnormal shadow on the chest X-ray was examined. A computed tomography scan of the chest performed during an intravenous drip infusion of contrast medium demonstrated an oval shaped area of density arising from the aortic arch and expanding obliquely downward toward the region of the aortopulmonary window (Fig. 5). Cineaortograms revealed an oval shaped aneurysm arising from the inferomedial surface of the aortic arch just distal to the branching of the left subclavian artery (Fig. 6). The aneurysm did not communicate with the pulmonary artery. The findings of computed tomography and cinearteriograms were concordant with findings of previously reported cases of ductus arteriosus aneurysm (7–14). The diagnosis of non-patent ductus arteriosus aneurysm was made.

Because of old age and poor cardiac performance, surgery was not performed. After admis-

![Fig. 5. Computed tomography scan of the chest during injection of contrast medium. An oval shaped density arises from the aortic arch and expands obliquely downward toward the aortopulmonary window. AAo, ascending aorta; DAo, descending aorta; L, pacemaker lead; lPA, left pulmonary artery; nPDAA, non-patent ductus arteriosus aneurysm.](image)


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sion, the patient was relieved of back pain and dyspnea.

**DISCUSSION**

It is now generally accepted that the two-dimensional echocardiogram is equal to or superior to angiographic techniques for the diagnosis of Ebstein's anomaly (6, 15–18). The attachment of the septal leaflet of the tricuspid valve well below the tricuspid valve annulus and displacement into the right ventricular cavity has been suggested as the most important findings in the diagnosis of Ebstein's anomaly (6, 15–18). Shina et al reported that all 41 patients with Ebstein's anomaly (except 5 with an absent septal leaflet) showed a displacement of more than 7 mm (range 7 to 50) (17). Inferior displacement of more than 8 mm/m² (indexed by body surface area) was exclusive to Ebstein's anomaly in comparison with a control group (17). In the present case, the body surface area was 1.56 mm² and this index was 22.4 mm/m².

The natural history of Ebstein's anomaly is variable but is compatible with long survival (4–6). Fifty percent of those diagnosed in infancy die early (2), whereas late survival is reported into the ninth decade (6). The present case was thought to be one of the latest survivals of Ebstein's anomaly. In general, significant associated cardiac defects lead to a worse prognosis (2). About 30 to 50% of patients with Ebstein's anomaly have a significant associated cardiac anomaly such as atrial septal defect, patent foramen ovale or patent ductus arteriosus (1–3). The present case had neither atrial septal defect nor patent foramen ovale, but had a non-patent ductus arteriosus aneurysm. This is the first report of the association of Ebstein's anomaly and ductus arteriosus aneurysm.

Various types of rhythm disorders have been documented in Ebstein's anomaly (1–3). Supraventricular tachycardia is a common feature of Ebstein's anomaly with a reported occurrence of 20–30% (2), but chronic bradycardiac atrial fibrillation is rare (1–3). This is the first report of Ebstein's anomaly combined with bradycardiac atrial fibrillation requiring pacemaker implantation.

Due to the variety of ductus arteriosus aneurysms, it is difficult to ascertain a definite pathogenesis of aneurysm. However, three
pathogenetic mechanisms have been suggested for the adult variety of ductus arteriosus aneurysm: 1) delayed spontaneous closure of a patent ductus with closure only at the pulmonary end; 2) trauma to a patient with persistence of the ductus; and 3) infective arteritis in a patient who has a patent ductus with subsequent spontaneous closure of the pulmonary arterial end (12–14). Because there was no history of chest trauma or sepsis in the present case, it was concluded that the first mechanism was probable. When complications such as rupture and dissection occur, most of the reported cases have a fatal outcome (9, 10, 13, 14). In the present case, surgery was not performed because the risks were felt to outweigh the benefits.

REFERENCES