INTRODUCTION

Ebstein’s anomaly (EA) is a rare congenital cardiac malformation of the tricuspid valve and the right ventricle occurring in less than 1% of all cases of congenital heart disease [1,2]. The natural history of EA is extremely variable, however, fewer than 5% of patients with EA survive beyond the age of 50 without surgical intervention [1,2].

In this paper, we report two unoperated cases of EA in adult patients over 50 years of age.

CASE REPORT

Patient 1: A 70-year-old Japanese woman, who had been diagnosed as EA, patent foramen ovale, tricuspid regurgitation (TR), and paroxysmal atrial fibrillation at the age of 26, was transferred to our hospital for syncpe. She had a history of cerebral embolism, maybe paradoxical, 4 times in her 30s and supraventricular tachyarrhythmias and/or heart failure several times in her 60s. Electrocardiography (ECG) showed atrial fibrillation with a wide QRS complex. After restoring sinus rhythm, ECG revealed a prolonged PQ interval, tall and broad P waves on II, III, aVF leads, and complete right bundle branch block but no delta wave. Chest X-ray showed severe cardiomegaly (CTR=70.0%) and decreased pulmonary vascularity. Transthoracic echocardiography demonstrated marked enlargement of the right atrium and a normal-sized left ventricle with good contraction. Apical displacement (25.3 mm/m² body surface area) of the septal leaflet of the tricuspid valve from the atrioventricular ring with tricuspid regurgitation in both patients. The former suddenly expired 20 months later after suffering from repetitive supraventricular tachyarrhythmias and/or heart failure, and the latter is alive with minimal signs of heart failure 12 months after the diagnosis of EA. Although the natural history of EA is extremely variable, these two patients are exceptional in that they tolerated EA well for over 50 years without any surgical intervention.

Summary: Ebstein’s anomaly (EA) is a rare congenital heart disease of the tricuspid valve, and less than 5% of patients with EA survive beyond the age of 50. We report two unoperated cases of EA in adult patients aged over 50 years. Two patients, a 70-year-old Japanese woman and a 59-year-old Chinese woman, were referred to us for tachyarrhythmias. Transthoracic echocardiography demonstrated apical displacement (>8 mm/m² body surface area) of the septal leaflet of the tricuspid valve from the atrioventricular ring with tricuspid regurgitation in both patients. The former suddenly expired 20 months later after suffering from repetitive supraventricular tachyarrhythmias and/or heart failure, and the latter is alive with minimal signs of heart failure 12 months after the diagnosis of EA. Although the natural history of EA is extremely variable, these two patients are exceptional in that they tolerated EA well for over 50 years without any surgical intervention.

Key words: Ebstein’s anomaly, congenital heart disease, tricuspid valve, tricuspid regurgitation, tricuspid septal leaflet, apical displacement of the tricuspid valve, echocardiography.
further examination and surgical intervention. Thereafter, the patient was conservatively managed, however she suffered from supraventricular tachyarrhythmias and/or heart failure which required hospitalization 5 times during the following 20 months before the patient died suddenly at her home. Autopsy was not permitted.

Patient 2: A 59-year-old Chinese woman was transferred to our hospital because of suddenly developed palpitation. Chest X-ray showed moderate cardiomegaly (CTR=61.5%) with normal pulmonary vascularity. ECG revealed supraventricular tachycardia with a
wide QRS complex. After restoring sinus rhythm, tall and broad P waves on II, III, aVF leads, and incomplete right bundle branch block without a delta wave were found on ECG. Transthoracic echocardiography demonstrated moderate dilatation of the right atrium and a normal-sized left ventricle with good contraction. Apical displacement (11.1 mm/m² body surface area) of the septal leaflet of the tricuspid valve from the atrioventricular ring was also detected. Doppler study showed moderate-to-severe TR but interatrial communication was not detected (Fig. 2). The patient refused further examination and surgical intervention. She is doing well with minimal signs of heart failure on diuretic and antiarrhythmic medications 12 months after the diagnosis of EA.

DISCUSSION

EA is a rare congenital heart disorder characterized by apical displacement of the proximal attachment of the septal and posterior leaflets of the tricuspid valve from the atrioventricular ring. In normal human hearts, the apical displacement of the septal and posterior leaflets is less than 8 mm/m² body surface area. Echocardiographic demonstration of an apical displacement of the tricuspid valve leaflet over 8 mm/m² is the most diagnostic predictor of EA [3].

The natural history of EA is extremely variable with a broad clinical spectrum from severe disease causing fetal or neonatal death to mild disease with survival to old age, depending on the severity of apical displacement and other associated heart disease [4]. Generally, of all neonates with EA, 20% to 40% do not survive 1 month, more than 50% die within the first 5 years, and less than 5% of patients with EA survive beyond the age of 50. In rare cases, patients with EA have been known to live over 70 years [2]. To our knowledge, the oldest living survivor was 87 years old as of 2002 [6].

Generally, patients with EA who survive into the adulthood are expected to have mild forms with good prognosis [4]. However, a follow-up study of 72 unoperated adult patients over 25 years have emphasized that many adult patients with EA may remain asymptomatic or minimally symptomatic for a long time but the prognosis may not always be good [5]. In the study, age at diagnosis less than 25 years, a CTR 0.65 or more, male sex and moderate or severe echocardiographic severity based on the septal leaflet attachment ratio have been indicated as risk factors for late cardiac-related death [5]. Generally, arrhythmia, particularly supraventricular arrhythmia, is the most common clinical feature at presentation in adult patients with EA but arrhythmia was not statistically associated with late cardiac-related death. However, paroxysmal supraventricular arrhythmia was a common cause of deterioration (class IV or III) in New York Heart Association (NYHA) functional class and prolonged recovery returning to functional class II or I after restoring sinus rhythm was a poor prognostic predictor. Similarly, when the functional class deterioration was not associated with an arrhythmic event, the risk of death was high [5]. These results suggest that surgical treatment should be contemplated in adult patients with EA who have clinical symptoms and signs such as a CTR 0.65 or more, moderate or severe echocardiographic severity, NYHA functional class III or IV and onset or progression of atrial or ventricular arrhythmias.

In conclusion, we reported two unoperated cases of EA of the tricuspid valve in adult patients over 50 years of age. Although the natural history of EA is extremely variable, the two patients described herein are exceptional patients who survived over the age of 50 years without any surgical intervention.

REFERENCES