When headaches are not neurological: an unusual presentation of Ebstein's anomaly

Introduction

This article reports the case of a 16-year-old male who presented acutely with a supraventricular tachycardia on a background of long-term migraine. This led to the diagnosis of a rare but important cardiac abnormality.

Discussion

The unique aspect of this case is the unusual presentation of undiagnosed congenital heart disease in a young adult.

Ebstein's anomaly is a congenital abnormality of the tricuspid valve. It occurs in 1 per 20 000 live births (Paranon and Acar, 2008) and is characterized by displacement of the septal tricuspid valve leaflet towards the apex of the heart (*Figure 4*). Part of the right ventricle becomes functionally part of the right atrium (i.e. 'atrialised'). The predominant problem is tricuspid regurgitation but obstruction can also occur. It is commonly associated with intracardiac shunts (e.g. atrial septal defect), accessory pathways and other congenital cardiac conditions.

Symptoms depend on the severity of the tricuspid valve lesion and other associated abnormalities. There may be cyanosis secondary to a right to left shunt, symptoms of right heart failure, palpitations as a result of supraventricular tachycardia or sudden death caused by ventricular arrhythmias.

Treatment options include medical therapy or radiofrequency ablation for arrhyth-

Dr C Cook *is ST2 in Medicine, St. Richards Hospital, Chichester PO19 6SE,* **Dr JP Carpenter** *is Specialist Registrar in Cardiology, Royal Brompton Hospital, London,* **Dr G Longcroft-Wheaton** *is Specialist Registrar in Gastroenterology and General Medicine, St. Richards Hospital, Chichester and* **Dr Y-K Wong** *is Consultant Cardiologist, St. Richards Hospital, Chichester*

Correspondence to: Dr C Cook

Case Report

A 16-year-old male was admitted following a loss of vision. Baseline observations demonstrated SpO2 80% on air, tachycardia of 180 beats per minute (bpm) and hypotension of 90/50 mmHg. He reported bilateral visual disturbance while sitting in bed together with breathlessness, chest discomfort and dizziness but had not had palpitation or syncope. He had been prescribed propranolol for recurrent migraines of increasing frequency with similar visual disturbances, nausea and vomiting but with no perceived benefit. He was otherwise well, a non-smoker who drank minimally and denied recreational drug use.

On examination, the jugular venous pressure was elevated but there were no other cardiorespiratory findings of note. Arterial blood gas analysis revealed profound hypoxia and a compensated metabolic acidosis. A 12-lead electrocardiogram demonstrated a supraventricular tachycardia with broad QRS (*Figure 1*). Carotid sinus massage and intravenous adenosine had no effect. Electrolyte levels were abnormal (potassium 2.7 mmol/litre, magnesium 0.45 mmol/litre, corrected calcium 1.60 mmol/litre) and a central venous line was inserted to allow electrolyte replacement. During this procedure, he spontaneously reverted to sinus rhythm at a rate of 70 bpm.

A repeat electrocardiogram showed sinus rhythm with normal axis but T wave inversion in the inferior leads and evidence of pre-excitation with a short PR interval and classical delta waves (*Figure 2*). As a result, Wolff–Parkinson–White syndrome was diagnosed. The chest radiograph showed an abnormal, globular cardiac silhouette (*Figure 3*).

Following return to sinus rhythm, the blood pressure and acid–base status normalized. Routine transthoracic echocardiography demonstrated Ebstein's anomaly with an associated atrial septal defect and small membranous ventricular septal defect.

Figure 1. 12-lead electrocardiogram showing supraventricular tachycardia with broad QRS complexes.

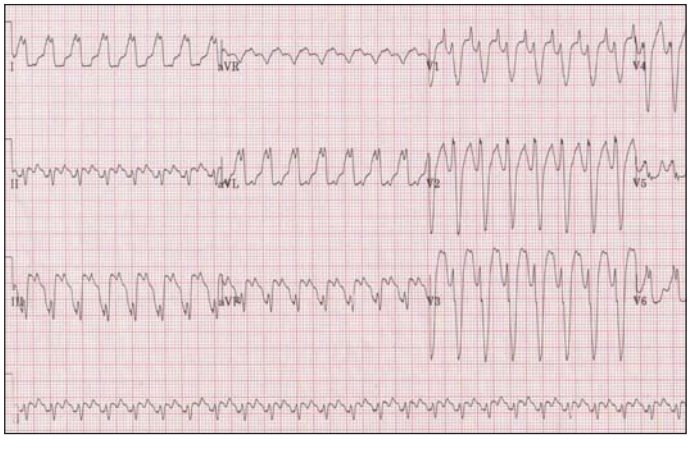
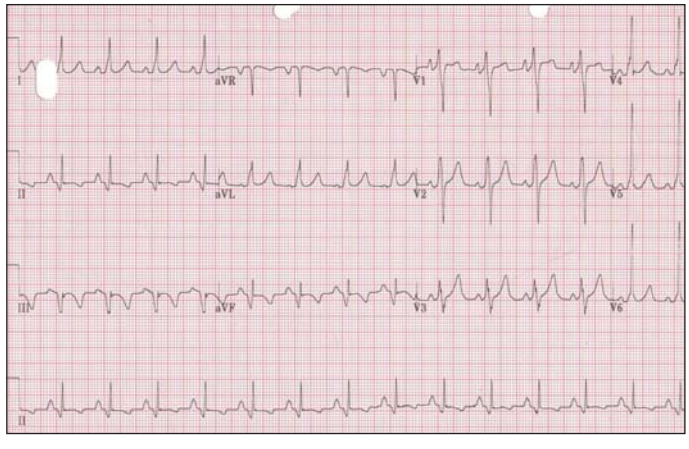


Figure 2. 12-lead electrocardiogram in sinus rhythm. Short PR interval and delta waves are most easily seen in lead I.



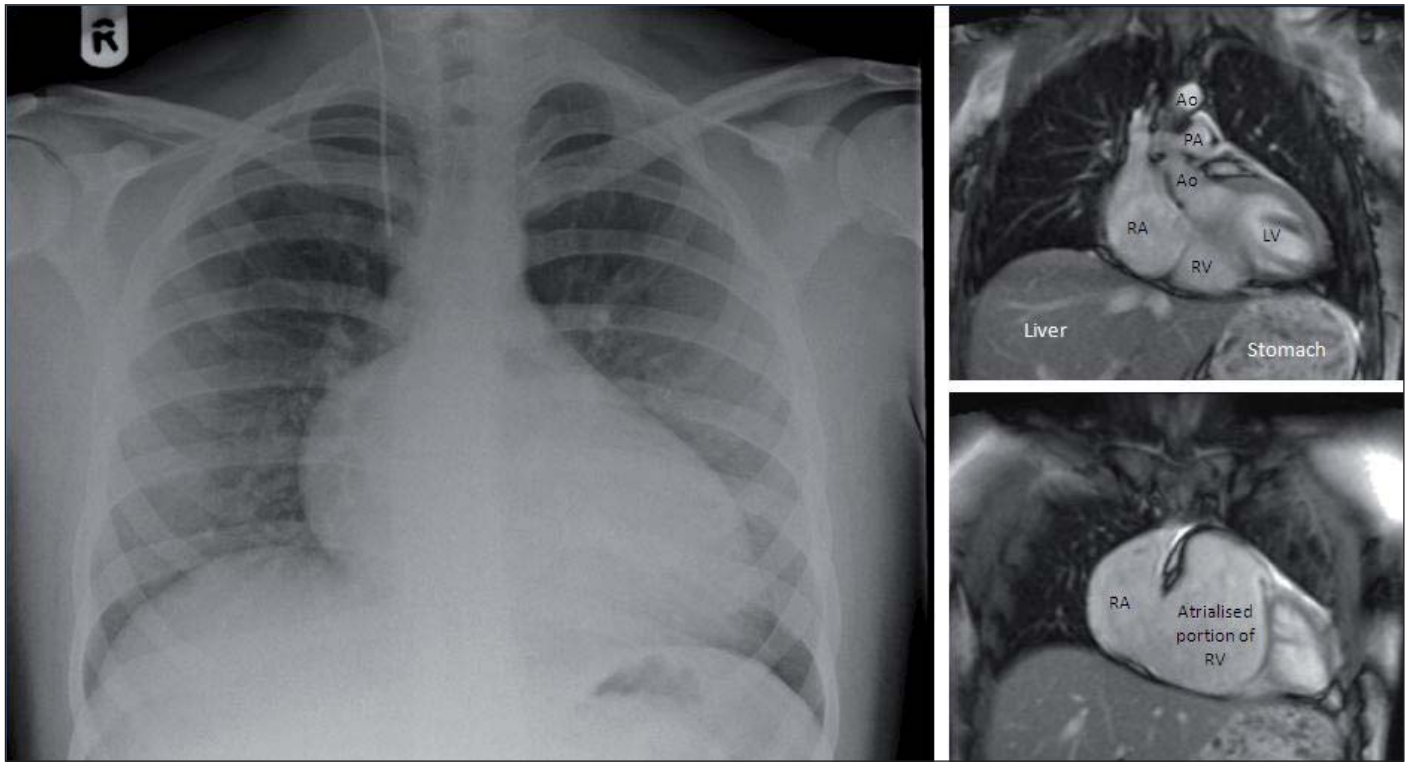


Figure 3. Chest radiograph and corresponding coronal magnetic resonance images demonstrating the cause of the enlarged cardiac silhouette. Ao= aorta; LV=left ventricle; PA= pulmonary artery; RA= right atrium; RV= right ventricle.

mias and surgical repair of the valve which can achieve near-complete restoration of anatomical and physiological function (Wu et al, 2007).

Conclusions

This case highlights the importance of considering cardiac pathology in patients presenting with migraine. The migrainous symptoms may have been caused by a low output state as a result of asymptomatic arrhythmia or even intracardiac shunting which has been well described as a cause of migraine (Del Sette et al, 1998). The significant electrolyte imbalance is likely to be the result of recurrent vomiting. Only full evaluation led to the diagnosis of Ebstein's anomaly. BJHM

Del Sette M, Angeli S, Leandri M, Ferriero G, Bruzzona GL, Finocchi C, Gandolfo C (1998) Migraine with aura and right–to–left shunt on transcranial Doppler: a case control study. *Cerebrovasc Dis* **8**(6): 327–30

Paranon S, Acar P (2008) Ebstein's anomaly of the tricuspid valve: from fetus to adult: congenital heart disease. *Heart* **94**(2): 237–43

Wu Q, Huang Z, Pan G, Wang L, Li L, Xue H (2007) Early and midterm results in anatomic repair of Ebstein anomaly. *J Thorac Cardiovasc Surg* **134:** 1438–42

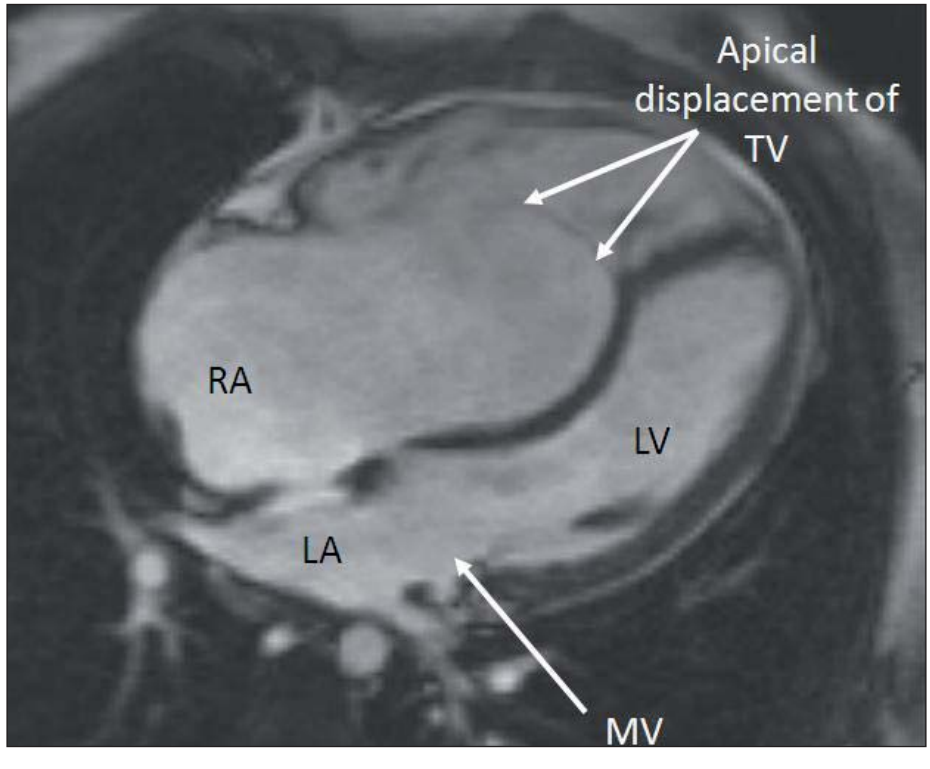


Figure 4. Cardiac magnetic resonance four-chamber view. There is marked apical displacement of the tricuspid valve (TV) typical of Ebstein's anomaly. LA= left atrium; LV= left ventricle; MV= mitral valve; RA= right atrium.