Ebstein's anomaly, Wolff-Parkinson-White syndrome and rheumatic mitral stenosis: role for combined electrophysiological and surgical management

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ABSTRACT

The coexistence of rheumatic mitral stenosis, Ebstein's anomaly and Wolff-Parkinson-White syndrome is an uncommon entity. To our knowledge, the successful management of this combination of lesions has not been previously described. We report a 23-year-old woman with the combination of these abnormalities. She was managed with preoperative electrophysiological study, followed by mitral valve replacement and Danielson's repair of tricuspid valve. The management issues involved are discussed in detail.

Fig. I 12-lead ECG shows short PR interval and delta waves suggestive of right free wall accessory pathway.

INTRODUCTION

The coexistence of Ebstein's anomaly (EA) with mitral stenosis (MS) has uncommonly been described.[1-7] This condition has profound haemodynamic significance. Additional presence of accessory atriventricular pathways, which are known to occur in up to 25% of patients with EA, poses a lot of specific management issues. To our knowledge, successful management of this combination of lesions has not been described to date. We report a young adult with this unusual combination of EA, Wolff-Parkinson-White (WPW) syndrome and rheumatic MS. She underwent hybrid management with presurgical electrophysiological (EP) study, and surgical repair of EA and coexisting MS successfully.

CASE REPORT

The patient was a 23-year-old woman who presented with a history of recurrent episodes of palpitation for a few years and dyspnoea on exertion for a few months. She gave a history suggestive of rheumatic fever in childhood. On examination, she was noted to have a regular pulse, with a rate of 86/minute, and raised jugular venous pressure with prominent a-wave. There was wide split S2, normal P2 and a low-pitched mid-diastolic murmur at the apex. Electrocardiography (ECG) showed normal sinus rhythm with preexcitation suggestive of right-sided accessory pathway (Fig. 1). Chest radiograph showed a cardiothoracic ratio of 65% with an enlarged right atrium and features of pulmonary venous hypertension. Transthoracic echocardiography revealed a mitral valve area of 0.9 cm^2 with a gradient of 22/12 mmHg across the mitral valve at a heart rate of 76/minute, mild aortic regurgitation, EA of the tricuspid valve, large atrial septal defect (ASD), trivial tricuspid stenosis, mild tricuspid regurgitation (TR) and mild pulmonary artery hypertension (PAH) (Fig. 2). The septal tricuspid leaflet was apically displaced by 21 mm with tethering to septum. Thickening of other leaflets and subvalvular chordal apparatus of tricuspid valve was also noted.

The echocardiographical findings were confirmed on cardiac catheterisation. The patient developed multiple episodes of narrow complex tachycardia

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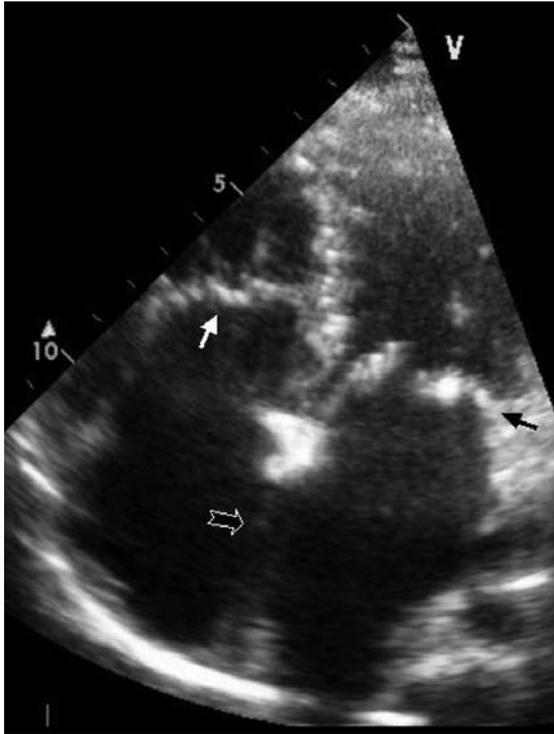Email: kknnamboodiri @sctimst.ac.in

Fig. 2 Transthoracic 2D echocardiography. Apical 4-chamber view shows Ebstein's anomaly of tricuspid valve (white arrow), large ostium secundum ASD (hollow arrow) and thickened mitral leaflets (black arrow).
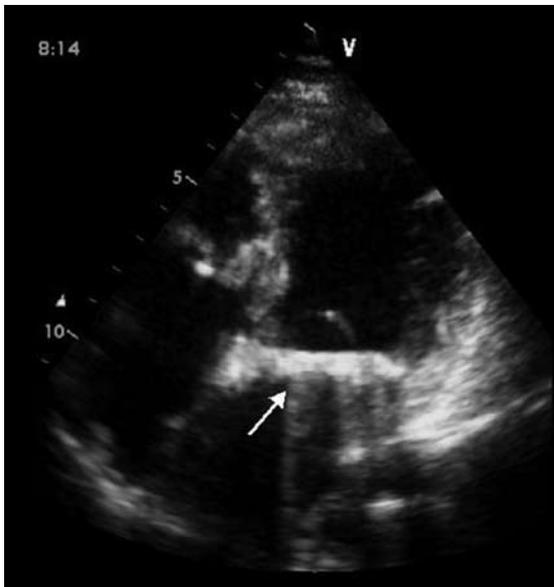

Fig. 3 Postoperative echocardiogram shows the mitral valve prosthesis (white arrow) and repaired tricuspid valve.

(heart rate 190–200/minute) during right heart catheterisation. Before surgery, the patient was subjected to an EP study to locate the abnormal pathway to facilitate radiofrequency ablation (RFA) prior to or during surgery. The EP study suggested the presence of multiple accessory pathways (AP) in the posterior aspect of tricuspid annulus and parahisian area. RFA was deferred in view of lack of ideal signals, presence of APs involving parahisian location and possibility of perioperative ablation. The patient was planned for mitral valve replacement and Danielson's repair for EA. The mitral valve was replaced with a 25-Chitra prosthetic valve, along with tricuspid valvotomy, closure of ASD and plication of atrialised segment of the right ventricle (Fig. 3). Amiodarone was given preoperatively and during one month of follow-up. The patient was asymptomatic and was not on any antiarrhythmics at 18 months follow-up. Though preexcitation of same pattern was evident in surface ECG, she did not have recurrence of palpitation.

DISCUSSION

EA is the most common congenital defect associated with the WPW syndrome. Manifest preexcitation and clinical tachycardia involving an AP, usually on the right side, can be present in up to 25% of patients.[8] Atrial flutter or fibrillation with accelerated conduction via AP may have serious physiological consequences in these patients, particularly in the presence of critical MS. There are many advantages in having an EP study in these patients preoperatively. First, APs, single or multiple, can be mapped, and can act as a guide to surgical ablation. Second, an attempt at ablation can be done prior to surgery, which can be aided by perioperative ablation in unsuccessful or partially successful cases. Third, catheter-based ablation can become more difficult in a postoperative altered anatomy. Fourth, failure to address APs with rapid antegrade conduction properties can lead to haemodynamic instability in the perioperative period in the presence of critically stenotic mitral valve if atrial arrhythmias develop. Finally, other cardiac arrhythmias, which are known to be associated with EA, may account for symptoms in a minority of patients, and can be identified and taken care of.

Precise localisation and ablation of APs in patients with EA can be quite challenging. Abnormal tricuspid valve anatomy, higher frequency of multiple pathways, catheter instability, fractionated electrograms at dysplastic annulus and propensity to have frequent atrial arrhythmias make the mapping and ablation difficult in these cases. The initial procedural success rate from various studies varies between 75% and 90% against more than 90% for APs in general.[9,10] Additional presence of MS makes the EP study more difficult. Rapid ventricular rates due to atrial arrhythmias induced during catheter manipulation or atrial pacing with anterograde conduction through the AP with low refractory period or re-entrant tachycardia mediated

by the AP can cause elevation of pulmonary venous pressure in presence of a stenotic mitral valve.

The haemodynamic consequences of EA are determined chiefly by the condition of tricuspid valve, the degree of haemodynamic impairment of the right ventricle and atrial rhythm. MS can predispose to atrial arrhythmias which, in presence of AP, can result in a rapid ventricular rate. The coexistent MS can alter the natural history of EA and cause atrial fibrillation, PAH and heart failure. In EA with TR, the development of PAH due to rheumatic MS should exacerbate preexisting TR. Rheumatic involvement of the tricuspid valve can also occur. In our patient, chordal thickening of the tricuspid valve by rheumatic involvement probably contributed to trivial tricuspid stenosis. However, a wide spectrum of deformity of the tricuspid valve can also occur in isolated EA.[11]

Surgical indication for concomitant correction of EA should be liberal if mitral valve surgery is planned. Our patient had no symptoms that could be attributed solely to EA, nor had she significant TR. MS can be managed in isolation without addressing the EA in such a setting, especially if the EA is mild. Balloon mitral valvotomy had been successfully performed in similar cases previously.[3] Bapat and Tendolkar preferred not to correct EA in such a case and did mitral valve replacement only,[7] while Ponoth et al treated with double valve replacement.[4] Open mitral valvotomy with plication of tricuspid valve has also been reported.[1] We preferred to do Danielson's repair of tricuspid valve and mitral valve replacement. Annular resection and resuturing are known to be successful in refractory arrhythmias mediated by AP. However, in our patient, this was not tried as APs also existed in the parahisian area. However, despite the presence of preexcitation on surface ECG, the patient remained free of palpitation 18 months postoperatively. Surgical repair of EA, without the aid of any surgical techniques for arrhythmia, is known to reduce the incidence of arrhythmias postoperatively, probably acting by the interruption of accessory pathways. In fact, less than one-sixth of patients continued to have postoperative symptomatic arrhythmias in this series.[12]

The combination of rheumatic MS, EA and WPW syndrome, and the management issues involved in it had not been described to date. We advocate preoperative EP study and ablation, if possible, in similar patients, though we could not ablate the AP in our patient due to parahisian location of the pathways.

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