A rare case report of WPW syndrome type B with non compaction of left ventricle, mitral valve prolapse and dilated cardiomyopathy

BALASUBRAMANIAM CHINNASAMY
Department of General Medicine,
COIMBATORE MEDICAL COLLEGE

Abstract:
Majority of the individual with Wolff-Parkinson-White syndrome (WPW) have normal heart, but may be associated with a number of cardiac pathology, especially congenital disease, in 7.5 to 17 percentage of cases. The combination of WPW with non-compaction of left ventricle, MVPS and DCM is rare situation. In this case, we are reporting 25yr old male patient, who presented with palpitation and was found to have this combination. The patient was successfully treated with radiofrequency ablation.

Keyword: Wolff-Parkinson-White syndrome, Mitral valve prolapse

INTRODUCTION:
WPW syndrome is a type of atrioventricular reentrant tachycardia. The incidence of WPW syndrome is between 0.1% and 0.3% of the general population. In most cases it is associated with a structurally and functionally normal heart, though occasionally can be associated with congenital heart disease such as Ebstein's anomaly (10%), MVPS (7%), DCM, HOCM(4%). Only 27 cases of accessory pathway-induced DCM have been reported thus far. We are presenting this case, a 25yr old male admitted for palpitation, found to be WPW syndrome. Combination of WPW syndrome with other cardiac anomaly is rare, combination of all these anomalies with WPW in single patient is very rare.

CASE REPORT:
A 25yr old male was admitted with the history of palpitation since 12yrs of age, which is sudden in onset and sudden termination, paroxysmal type, more on exertion. No history of syncope, chest pain, breathlessness and failure features. Past h/o and family h/o not relevant. O/E patient conscious, oriented, not dyspnoeic, no pedal edema, BP 110/70 and pulse 88/min. Systemic examination CVS: SI,S2 heard, no murmur, RS: clear, other systems normal. ECG taken immediately which showed:
ECG

NSR, HR 70/min, left axis deviation, short PR interval. Broad QRS complex, negative delta waves present in lead V1, aVR.

Positive delta waves in II, aVF. Indicated that WPW syndrome type B and accessory pathway in RV free wall. ECHO was done which showed,

ECHO

LA/LV dilated, Prolapse of AML, Prominent trabeculation of LV, Borderline systolic dysfunction, Global hypokinesia of LV, LVEF 55%, Doppler velocity normal, pericardium normal.

Routine blood investigations were done, which showed normal range. Then electrophysiological study was done to know the number and location of accessory pathways.

Since our patient is young and symptomatic, more risk for tachyarrhythmias and sudden death, we planned for radiofrequency ablation for permanent cure. Since facilities for electrophysiological study and radiofrequency ablation were not available in our institution, we referred this patient to JIPMER for further management. Pt was treated using long sheath approach 4mm tip St jude therapy catheter was used for mapping and ablation. Early activation was mapped to 10’0 clock on the tricuspid annulus. Long sheath was used for stability. RF ablation resulted in loss of preexcitation. Patient is under follow up in our institution.

DISCUSSION:

WPW syndrome is a congenital abnormality associated with supraventricular tachycardia. It involves an activation of the ventricles that occur earlier than expected, called preexcitation, which occurs because of conduction of an atrial impulse not by means of the normal conduction system, but via an extra atrioventricular (AV) muscular connection, termed an accessory pathway, that bypasses the AV node. This accessory electrical pathway is present in approximately 1.5 people per 1,000 people. It runs in families in less than 1% of cases.

WPW syndrome is found in persons of all ages, from those in fetal and neonatal age groups to elderly individuals. Prevalence decreases with age because of loss of preexcitation. In patients with abnormal ECG findings indicative of WPW syndrome, the frequency of SVT paroxysms increases from 10% in people aged 20-39 years to 36% in people older than 60 years.
From 7% to 20% of patients with WPW syndrome have other accompanying congenital abnormalities, the most common being Ebstein’s anomaly, which is associated with single or multiple right-sided APs. Other rare associations include hypertrophic cardiomyopathy, dilated cardiomyopathy, mitral valve prolapse, atrial septal defect, ventricular septal defect, transposition of the great vessels, coarctation of the aorta, dextrocardia, coronary sinus diverticula, right and left atrial aneurysms, cardiac rhabdomyomas, Marfan’s syndrome, and Friedreich’s ataxia.

In our case WPW syndrome is associated with dilated cardiomyopathy, mitral valve prolapse, non compaction of left ventricle. Only 27 cases of accessory pathway-induced DCM have been reported thus far. Recently, several investigators have demonstrated reduced systolic LV function in patients with right- sided septal and paraseptal accessory pathways. A lower LV ejection fraction was noted in 56% of patients with septal and paraseptal accessory pathways. In a study by Know et al, systolic function was reduced in all patients with septal pathways, compared to patients with left or right-sided free wall pathways. An important clinical finding is that cardiac function improves and normalizes in patients with LV dysfunction or DCM, following loss of ventricular preexcitation, either spontaneously or due to cardiac catheter ablation or medical therapy. The prognosis of accessory pathway-induced DCM is excellent. There are a number of reports of a definite association between preexcitation atrioventricular pathways and MVPS. The relation with the Wolff-Parkinson-White syndrome was first reported by Gallagher et al in 1975 with seven cases of MVPS among 68 patients with the WPW syndrome and attacks of arrhythmia that were sufficiently frequent or of such severity as to be disabling or life endangering.

Isolated noncompaction of the left ventricular myocardium (INVM) is a rare, congenital abnormality resulting from an arrest in normal endomycocardial embryogenesis. Although INVM is a congenital myocardial disorder, the onset of symptoms is frequently delayed until adulthood. The origin of the high incidence of WPW syndrome in INVM remains unclear. The ECG findings in INVM are related to WPW syndrome type B because defects of the annulus fibrosus in cardiac embryogenesis explain the subendocardial position of the accessory atrioventricular connections around the tricuspid valve.

Reference: