

Wilson's Disease with Wolff-Parkinson-White Syndrome: Case report

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Abstract

A cardiac involvement in Wilson's disease includes left ventricular hypertrophy, severe atherosclerosis, sudden cardiac death, and various arrhythmias. In this report, a case with Wilson's disease and Wolff-Parkinson-White syndrome is discussed with the relevant literature. A 22-year-old male patient presented to the hospital complaining of chest pain. The patient had the diagnosis of Wilson's disease. He had short periods of palpitations for 2 years but no tachycardic period was detected in his 24-hour electrocardiogram holter monitoring. An electrocardiogram showed a shortened PR interval and prominent delta waves supporting the diagnosis of Wolff-Parkinson-White syndrome. The patient was hospitalized and subsequently transferred to electrophysiologic mapping. The accessory pathway was eliminated with radiofrequency ablation catheter. Patients with Wilson's disease who have palpitations should be evaluated for possible arrhythmias and electrocardiography should be performed. In this report, a case with Wilson's disease and Wolff-Parkinson-White syndrome is discussed with the relevant literatures.

Key Words: Disorder Of Copper Metabolism; Preexcitation Syndrome; Genetic Diseases.

Wilson Hastalığıyla Birlikte Gösteren Wolf-Parkinson-White Sendromu: Olgu sunumu

Özet

Wilson hastalığının kardiyak tutulumunda sol ventrikül hipertrofisi, aterosklerozis, ani kardiyak ölüm ve çeşitli aritmiler görülebilir. Bu yazıda Wolff-Parkinson-White sendromunun eşlik ettiği Wilson hastalığı literatürler ışığında tartışılmıştır. Wilson hastalığı tanısı olan 22 yaşında erkek hasta göğüs ağrısı şikâyeti ile hastaneye başvurdu. 2 yıldır kısa süreli çarpıntı şikâyetleri olan hastanın 4 saatlik elektrokardiyografik holter incelemesinde herhangi bir taşikardi saptanmamış. Hastanın elektrokardiyografisinde Wolff-Parkinson-White sendromunu destekleyen kısa PR aralığı ve delta dalgası bulunmaktaydı. Hasta hastaneye yatırıldı; sonrasında elektrofizyolojik haritalama yapıldı ve radyofrekans ablasyon yöntemi ile aksesuar yolak sonlandırıldı. Çarpıntı şikâyeti ve Wilson hastalığı olan hastalar olası aritmiler yönünden değerlendirilmeli ve bu hastaların elektrokardiyografileri aritmi açısından incelenmelidir. Bu yazıda Wilson hastalığı ve Wolff-Parkinson-White sendromu olan bir vaka ilgili literatürler eşliğinde sunulmuştur.

Anahtar Kelimeler: Bakır Metabolizma Bozukluğu; Preeksitasyon Sendromu; Genetik Hastalıklar.

INTRODUCTION

Wilson's disease (WD) is an autosomal recessive inherited disorder of copper metabolism, resulting in pathological accumulation of copper in many organs and tissues (1). The worldwide prevalence is about 1 in 30,000, which may vary by population (2). The culprit gene is ATP7B which is localized on the long arm of chromosome 13 and codes for a copper-transporting P-type ATPase (3). The hallmarks of the disease are the presence of liver disease, neurologic symptoms, and Kayser-Fleischer corneal rings (1). There are only few studies evaluating cardiac manifestations of WD (4,5). Cardiac involvement of WD includes cardiac hypertrophy, interstitial fibrosis, atrioventricular

nodal degeneration, severe atherosclerosis, sudden cardiac death and various arrhythmias (4). Wolff-Parkinson-White syndrome (WPWS) is the second most common form of paroxysmal supraventricular tachycardia. The prevalence of WPWS is 1.5 to 3.1 per 1,000 persons in Western countries. There is an accessory bundle of muscle tissue, which connects the atrium to the ventricle bypassing the atrioventricular node (6). Here we report first case of WD with WPWS.

CASE REPORT

A 22-year-old male patient presented to the cardiology polyclinic complaining of chest pain which occurred immediately after a sportive

accident. The patient had the diagnosis of WD since he was 12-years-old and had been taking oral penicillamine (1200mg/d) and zinc (600 mg/d). The patient had WD since he was 12 years-old and he has been taking 1200 mg penicillamine and 600 mg zinc daily. He had short periods of palpitations for 2 years but no tachycardic period was detected in his 24-hour electrocardiogram Holter monitoring. None of his siblings had a similar illness. General physical examination at the time of admission was normal except for mild hepatomegaly. Neurological examination revealed normal findings. The biochemical investigations including liver enzymes were in normal ranges, but indirect bilirubin levels were as high as 1.4 mg/dl (normal range 0.1-0.9 mg/dl). An electrocardiogram showed a shortened PR interval and prominent delta waves supporting the diagnosis of WPWS (Figure 1). The echocardiographic examination was normal. Genetic testing could not be performed because of technical issues. The patient was hospitalized and subsequently transferred to electrophysiologic mapping. Right ventricular posteroseptal accessory pathway was identified. The accessory pathway was eliminated with radiofrequency ablation catheter and delta wave had disappeared. A few minutes later, conduction from the deep accessory pathway returned and delta wave was seen again. The procedure was postponed because of the deeper localization of the accessory pathway. We planned to retry ablation with an internally cooled intramural needle ablation catheter soon and discharged the patient on 5 mg/day bisoprolol.

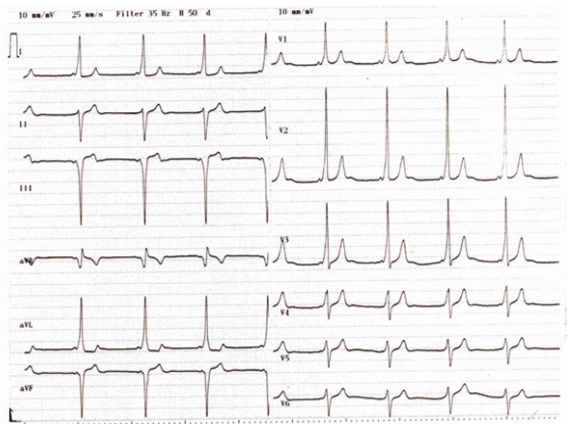


Figure 1. Electrocardiogram of the patient showing preexcitation characterized with short PR interval and delta waves.

DISCUSSION

Cardiac abnormalities in WD have been associated with excessive systemic accumulation of various metals (copper, iron, lead, etc) (4). Cardiac hypertrophy was detected in 5 out of 9 cases (55%) in an autopsy series of patients with WD. Two of the 9 patients had died suddenly, presumably secondary to an arrhythmia (5).

In a series of 53 patients with WD, Kuan had reported that 34% of the patients had electrocardiographic abnormalities and 19% had orthostatic hypotension (7).

During a ten-year follow-up, two cardiac deaths (0.6% per patient-year) had occurred. One patient died of ventricular fibrillation. Twenty-four-hour electrocardiogram holter monitoring detected electrocardiographic abnormalities in 42% of the patients with WD (4). The most frequent findings were supraventricular tachycardias and frequent supraventricular ectopic beats. Early repolarization, ST depression, T wave inversion, atrial fibrillation, sinoatrial block, Mobitz type 1 atrioventricular block and sudden death due to ventricular fibrillation were also defined (4,7).

WPWS as a cause of sudden cardiac death, presumably as a result of rapidly conducting atrial fibrillation, is well recognized. Rapid heart rates whether retrograde or antegrade, if sustained might lead to heart failure or even ventricular tachycardia and sudden cardiac death. WPWS can be a sporadic disease or genetical inherited disorder like WD. Mutations in the PRKAG2 gene located on chromosome 7q36 which encodes the gamma2 regulatory subunit of AMP-activated protein kinase had been shown to cause autosomal dominant WPWS associated with hypertrophic cardiomyopathy (6). A novel mutation in PRKAG2 causing WPWS and conduction system disease with onset in childhood and the absence of cardiac hypertrophy has also been reported (8). The accepted form of treatment in patients with familial WPWS is ablation of the accessory bundle.

WPWS occurring in association with different genetic disorders like tuberous sclerosis have been reported but concurrent WD has not been

described before (9). This represents the first reported case in the literature that WPWS developed as a disease process of WD. Also it may be an independent clinical manifestation irrelevant to WD because the patient presented above did not have cardiac hypertrophy or atrial fibrillation. Besides, none of his relatives had a history of sudden death or preexcitation syndrome. So it is unclear that WPWS developed as a disease process of WD or is an independent clinical manifestation irrelevant to WD.

In conclusion, patients with WD who have palpitations should be evaluated for possible arrhythmias and electrocardiography should be performed.

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