Case Report

PAROXYSMAL ORTHOSTATIC TACHYCARDIA SYNDROME (POTS) WITH CO-EXISTING CHRONIC FATIGUE SYNDROME: A REVIEW OF THREE CASES AND DISCUSSION

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ABSTRACT

Orthostatic intolerance appears in many guises including overt dysautonomia, vasovagal syncope and orthostatic tachycardia. We present details of three patients referred to our syncope clinic, who satisfied the chronic fatigue criteria of the centre for disease control and prevention. Head-up tilt testing induced significant hypotension and increased heart rate in all three patients, consistent with the clinical and autonomic manifestations of postural orthostatic tachycardia syndrome. We report on the emerging evidence available which does suggest a direct relationship between these two syndromes.

KEY WORDS: POTS, CFS, Vasovagal syncope.

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INTRODUCTION

Patients with chronic fatigue syndrome sometimes show features of postural orthostatic tachycardia during tilt table testing. Conversely, patients with POTS often have fatigue as a prominent feature. Symptoms may be provoked using GTN. We present details of three patients, referred to our syncope clinic for investigation of syncope and presyncope, who showed features of both Chronic fatigue and Orthostatic tachycardia syndromes.

Case Report 1: A 41 year old man who works as a mechanic for one of the national motoring organisations was admitted to the medical assessment unit of our District general hospital in June 2004 having had a syncopal episode. He also complained of feeing unwell, tiredness, blurring of vision, sweating and intermittent palpitations. There were no reports of incontinence or tongue-biting. He had just completed his 6th day of long shifts at work. He had a similar episode of collapse about 10 years previously while driving, having had a preceding viral illness. At that time PAF was diagnosed and he was started on â-blockers. He was unable to tolerate this and it was subsequently discontinued. It is worth noting that he has an uncle who presented with similar symptoms.

On examination his general observations were normal. His ECG showed AF/Flutter with a normal axis. All his routine bloods including TFTs were normal. His Holter tape showed sinus rhythm with a few dropped beats. It was thought that he probably had vaso-vagal syncope and was referred for tilt testing. His initial Tilt test showed significant resting tachycardia throughout the tilt associated with significant hypertension. It was thought that this resting tachycardia may have been indicative of neuro- cardiogenic syncope, so it was decided to repeat the tilt test with

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GTN provocation. On further questioning he admitted that his dietary salt intake was very low and that his fluid intake was also probably inadequate. In the meantime he was advised to increase his salt and fluid intake. When he presented for his repeat tilt test with GTN provocation, he reported feeling a lot better after increasing his dietary salt and fluid intake. The repeat tilt test demonstrated an even more marked resting tachycardia during tilt, with a further increase in heart rate after 400mcg of sublingual GTN up to a maximum of 142 beats/ min. This was associated with symptoms of dizziness and disorientation which were similar to his usual pre- syncopal symptoms. After lowering to the supine position, his heart rate returned to baseline levels after two minutes. These findings were suggestive of POTS. On further questioning the patient described features symptoms which included tiredness and lethargy. These features are also described by patients with CFS which is often attributed to a disturbed sleep pattern. It emerged that the patient works in a shift pattern over the last 7-8 years has completely disturbed his sleep. The management of this patient included increasing his dietary salt and fluid intake and, after liaison with his employer, his shift pattern was modified.

Case Report 2: A 21 year old woman was referred by the cardiologists to the syncope clinic with a history of having had recurrent syncopal episodes since the age of eight years. Most of these episodes occurred while standing although a few had occurred while either sitting or lying down. The episodes were preceded by dizziness, blurred vision, and sometimes tinnitus after which she blacks out. She gave a history of feeling very lethargic and listless most of the time. She also had Asthma for which she was on Becotide and Salbutamol inhalers. Her 24 hours ECG showed sinus rhythm, but with a rate ranging from 51 to 167 beats per minute. Her tilt test showed persistent resting tachycardia during tilt, which returned to the baseline level immediately after lowering to the supine position. These features were suggestive of neurocardiogenic syncope, it was arranged for her to have a repeat tilt test with GTN provocation. The results of her tilt test showed a significant tachycardia after administering 400 mcg. Of GTN up to a maximum of 137 beats per minute. This was associated with feeling sweaty and palpitations, and two minutes later with a heart rate of 134 beats per minute and a corresponding blood pressure of 106/50 mmHg, she felt even more sweaty and dizzy and became pre-syncopal. Both her heart rate and blood pressure returned to baseline levels immediately after lowering to supine position. With a diagnosis of postural orthostatic tachycardia in mind, with her history of underlying asthma, she could not benefit from beta blockers. She did admit to not adding any salt to her food, so she was advised to increase her dietary salt and repeated the tilt test in two months time. Since increasing her dietary salt she reported a significant improvement in her symptoms. Her dizzy spells were also far less frequent and she managed by sitting down as soon as she started to feel symptomatic. Although her tilt test again demonstrated a resting tachycardia during tilt, it was less marked than on previous occasions. She was advised to continue with her current management and discharged from our clinic.

Case Report 3: A 43 year old woman was referred to our syncope clinic with a history of recurrent syncopal episode for the past 8 months. These episodes were precede by feeling hot, sweaty and dizzy, and this was soon followed by a blackout which could be avoided by either sitting or lying down. She usually recovered within minutes and most of these attacks occurred while standing. She also gave a history of tiredness, general body ache and disturbed sleep. Her 24 hours ECG showed nothing significant. Her tilt test showed a slight and relative increase in her resting heart rate during the tilt, which soon settled down after lowering to the supine position after 35 minutes. The tilt test was inconclusive, but in view of the slight tachycardia, it was arranged for her to have a repeat tilt test with GTN provoca-

tion. Within two minutes of administering GTN her pulse rate increased to 128 beats per minute, at which stage she became pre-syncopal. She was lowered to the supine position and within three minutes her heart rate had returned to baseline levels. These findings were suggestive of postural orthostatic tachycardia syndrome. After discussing treatment options with her, and there being no contraindications, she was started on the beta blocker Metoprolol. She was also advised about increasing her dietary salt and fluid intake.

DISCUSSION

At the beginning of nineteenth century, physicians reported patients suffering from fatigue and poor exercise tolerance which occurred without an obvious cause. Some of the first reports, by DaCosta were on soldiers who fought in the American civil war and he coined the term "irritable heart syndrome" and "soldier's heart".[1]

POTS is operationally defined by the presence of symptoms of orthostatic intolerance associated with an increase in sinus heart rate of >30 beats/min. Or to a rate of 120 beats/minutes, during the first 10 minutes of tilt table testing.[2] The Centre for disease control criteria for CFS include chronic debilitating fatigue lasting for >6 months associated with cognitive difficulties, pharyngitis, tender lymphadenopathy, muscle pain, joint pain, headache, sleep disturbances and post exercise malaise unexplained by other illness.[3,4] Five previous studies have shown that patients with CFS sometimes have findings of POTS during tilt table testing.[5,6] Conversely, patients with POTS often have fatigue as a prominent feature.[2,5,6] Vagal withdrawal with intact vasoactive baroreflexes and sympathoexitation may contribute to vasomotor instability and orthostatic intolerance in these patients. Symptoms can be provoked by using GTN.[7]

The emergence of tilt testing as a reliable method for provoking these periods of autonomic decompensation not only provides a useful diagnostic tool but has also allowed for a much better understanding of the pathophysiology of these disorders. The suggested mechanisms of POTS[7-9] have included hypovolemia, excessive venous pooling while standing, loss of adequate vascular tone in the lower extremities, beta – adrenergic hypersensitivity of cardiac receptors.[5] Attenuated post-viral panautonomic neuropathy has also been proposed as a cause of dysautonomia. Our patient gives a history of a viral illness prior to his first episode of illness 10 years ago.[7] Some patients show a familial predisposition. Our patient also gives the history of an affected uncle. The defective gene causes a dysfunction in a nor epinephrine transporter protein, producing excess serum nor epinephrine levels.[10]

The management of POTS[7,11,12] includes an increase of salt and fluid intake. This seemed to have resulted in a significant improvement in our patients also. Patients are encouraged to sleep with the head end of the bed slightly elevated, to avoid extreme heat, dehydration and excess alcohol consumption. Elastic support hose (which is waist high and provides 30 mmHg ankle pressure) has been used with success. Drugs like Fludrocortisone, Beta blockers; Clonidine, Erythropoetin and Venlafaxine (SSRI) have been tried with moderate success.

The management of CFS involves combined behaviour therapy (CBT) and a major part of it is establishing a sleep routine which was done in our patients as well as graded exercise therapy. POTS and CFS are potentially recognisable and treatable disorders and evidence of a relationship between the two is emerging.[4,6] The importance goes beyond the number of people it affects, as it may cause substantial disability among young, otherwise healthy individuals. Therapies directed at correcting autonomic imbalance can often relieve the severity of symptoms. Greater efforts will be necessary to better understand these syndromes and their various subtypes and provide therapies that will help this group of highly symptomatic patients return to normal life.

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