ORTHOSTATIC INTOLERANCE: POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME WITH OVERLAPPING VASOVAGAL SYNCOPE

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SUMMARY – A 28-year-old female with a history of situational syncope and a new-onset right sided hemiparesis is described. Tilt-up table test revealed the postural orthostatic tachycardia syndrome followed by vasovagal syncope. Neurological and internal medicine tests showed no particular disorders. The patient underwent autonomic physical training and the tilt-up test performed three months later showed improvement of the autonomic system in terms of lower heart beat rate of the postural orthostatic tachycardia syndrome and longer duration of the test. This case report describes longstanding idiopathic dysautonomia that can be improved by nonpharmacological treatment, while reminding that this medical condition may also be the cause of syncope.

Key words: *Postural orthostatic tachycardia syndrome; Syncope; Dysautonomia*

Introduction

Postural orthostatic tachycardia syndrome (POTS) is observed predominantly in younger women and is defined as a sustained heart rate increase of ≥30 bpm or an increase to ≥120 bpm within the first 10 min of orthostasis associated with symptoms of orthostatic intolerance and without significant orthostatic hypotension. A higher prevalence of vasovagal syncope has been observed among patients with POTS. There are no accurate epidemiological studies regarding this syndrome. Its etiology is believed to be multifactorial with a number of possible underlying mechanisms and overlapping syndromes[1,2]. Here we present a 28-year-old female with a history of typical situational syncope and a new-onset right-sided hemiparesis.

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Case Report

A 28-year-old female was hospitalized at neurology department because of typical vasovagal syncope (prodromes occurred in a crowded warm place) followed by new-onset unilateral right-sided hemiparesis of the lower arm and leg. While unconscious, neither physical movements nor post-syncope diuresis/stool were observed in our patient. Medical history revealed recurrent syncopes in provocative surroundings since puberty, without any neurological disturbances recorded. Other patient history was unremarkable. Syncopes and symptoms such as lightheadedness and palpitations have transitorily worsened after each of the two normal pregnancies. The patient is current active smoker and had been using birth control pills for the past four months. She denied infection of any kind or insect bites recent to this episode.

Methods

Physical examination revealed intact cardiopulmonary finding, but showed right-sided neurologi-

cal performance to be weaker. Neurological testing (CT, MR, EEG, duplex Doppler, EMNG, CSF) showed no pathology. Laboratory findings (RBC, WBC, coagulation parameters, Fe, UIBC, TIBC, Fe saturation, thyroid hormones, HbA_{1c}, ESR, CRP, electrolytes, blood urea nitrogen, creatinine, aminotransferases, urine) were normal. Cardiac cause of the syncope was evaluated; there was no orthostatic hypotension, ECG was unremarkable as well as transthoracic echocardiography. Head up tilt table test revealed POTS followed by vasovagal syncope in the 25th minute of the test. Since then, the patient has been performing orthostatic workout regularly, taking care of daily fluid and salt intake, and has been undergoing physical therapy for three months. The following head up tilt table test showed no trace of hemiparesis and it was positive in the 48th minute of the test.

During the first tilt-up test, the heart beat rate was approximately 135-145/bpm with concomitant palpitations, at 95/70 mm Hg. Vasovagal syncope occurred in the 25th min at 45/bpm and blood pressure of 50 mm Hg palpatory (Fig. 1a). After three months of autonomic physical training, the second tilt-up test showed improvement in terms of lower heart beat rate and longer duration of the test. During the second tilt-up test, the heart beat was approximately 125/bpm with blood pressure of 90/75 mm Hg, while the patient was asymptomatic. In the 43rd minute, prodromes occurred at 140/bpm. In the 47th minute, a rapid decrease of pulse was noted, a minute later followed by vasovagal syncope at 52/bpm and blood pressure of 60 mm Hg palpatory (Fig. 1b). Both tilt-up tests were performed according to the 2009 Syncope Guidelines, without medical provocation[3].

Discussion

This is an interesting case presenting dysautonomia of an untraceable cause. Our patient's dysautonomia manifested as transitory hemiparesis and orthostatic impairment presenting as POTS with overlapping vasovagal syncope.

POTS etiology is still unknown and is supposed to be multifactorial, in the sense of sympathetic hypersensitivity, venous and arterial dilatation mechanisms at the microcirculatory level[4-7]. POTS is part of a wide spectrum of disorders that exhibit autonomic dysfunction. These include severe orthostatic hypotension in the presence of autonomic neuropathy and vasovagal syncope in the absence of other evidence of autonomic dysfunction. POTS manifests as an excessive orthostatic tachycardia without significant orthostatic hypotension in patients without overt autonomic neuropathy. It is associated with numerous other symptoms such as exercise intolerance, palpitations, weakness, and lightheadedness; most of these symptoms are also autonomically mediated[8].

Our patient's clinical impairment was improved by nonpharmacological maneuvers, proved by better physical performance (absence of no-tilt induced syncope and no manifest hemiparesis), objectified by longer maintaining of orthostatic stability at the recommended duration of the tilt up table test in persons with POTS[9]. Here we considered orthostatic home training and care of daily fluid and salt intake as sufficient therapy for this neurocardiovascular condition. This case report can be perceived as a contribution to the overall knowledge of POTS, as it is a relatively new medical term, and as a reminder to consider this

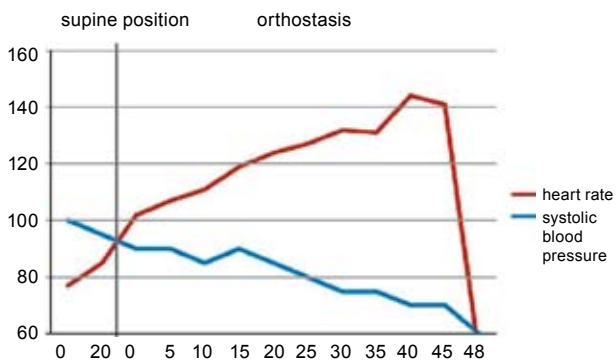

Fig. 1a. Tilt up table test 1.

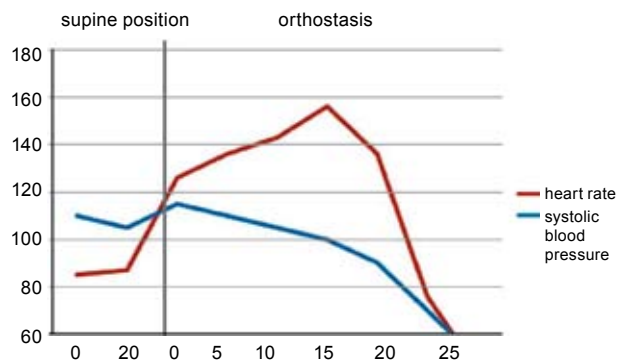

Fig. 1b. Tilt up table test 2.

condition when evaluating the potential causes of syncope.

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Sažetak

ORTOSTATSKA INTOLERANCIJA: SINDROM POSTURALNE ORTOSTATSKE TAHIKARDIJE S VAZOVAGALNOM SINKOPOM

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Prikazuje se 28-godišnja bolesnica s anamnezom višegodišnje situacijske sinkope i novonastalom desnostranom hemiparezom. Učinjen *tilt-up table* testom utvrđen je sindrom posturalne ortostatske tahikardije (engl. POTS, *postural orthostatic tachycardia syndrome*), nakon kojeg je uslijedila vazovagalna sinkopa. Neurološkom i internističkom obradom nije utvrđen eventualan drugi uzrok sinkope. Tijekom slijedeća tri mjeseca bolesnica je u kućnim uvjetima provodila ortostatske vježbe autonomnog sustava, nakon čega je učinjen kontrolni *tilt-up table* test kojim je objektivizirano poboljšanje statusa autonomnog sustava u smislu nižih vrijednosti frekvencije tahikardije POTS-a i duljeg trajanja testa, odnosno održavanja ortostaze. Ovaj slučaj govori o višegodišnjoj idiopatskoj disautonomiji na koju je moguće utjecati nefarmakološkim metodama te je ujedno i podsjetnik na jedan od mogućih uzroka sinkope.

Ključne riječi: *Sindrom posturalne ortostatske tahikardije; Sinkopa; Disautonomija*