J Med Sci 2010;30(3):127-129
http://jms.ndmctsgh.edu.tw/3003127.pdf

Idiopathic Vagal Palsy Complicated with Orthostatic Tachycardia

Yu-Hua Lai, Jiann-Chyun Lin, and Chun-An Cheng*

Department of Neurology, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan, Republic of China

Orthostatic tachycardia is a condition of orthostatic intolerance with disproportionate increases in heart rate. We describe a case of a 30-year-old man who presented with vagal palsy followed by orthostatic intolerance. The head-up tilt test demonstrated an increase in heart rate from 72 to 104 beats/min and a decrease in cerebral blood flow velocity from 60 to 27 cm/s at the right middle cerebral artery. Heart rate variability during deep breathing and the Valsalva ratio indicated parasympathetic impairment. This case confirmed that parasympathetic dysfunction with unopposed sympathetic activity can lead to orthostatic tachycardia. Hence, the autonomic function test in patients with idiopathic vagal palsy should be carefully evaluated to detect this rare complication.

Key words: orthostatic tachycardia, vagal palsy, parasympathetic dysfunction

INTRODUCTION

Orthostatic intolerance is defined as the development of symptom during upright standing and relieved by recumbency. Orthostatic tachycardia is a condition of orthostatic intolerance with disproportionate increases in heart rate.[1] Common clinical manifestations of orthostatic tachycardia include lightheadedness, dizziness, palpitations, nausea, difficulty breathing and fatigue. Vagal palsy is characterized by nasal speech, dysphagia and hoarseness. In addition to motor divisions, the vagus nerve also contains parasymapathetic fibers, which can control the heart rate through the baroreflex. It is easy to overlook and underestimate the cardiovascular consequence in vagal palsy. We present a case of a 30-year-old man with idiopathic vagal palsy complicated with orthostatic tachycardia. The relationship of orthostatic tachycardia and vagal palsy is also discussed.

CASE REPORT

A 30-year-old man developed sudden onset of slurred speech with nasal timbre, difficulty in swallowing, hoarseness, and regurgitation through the nose for 2 days. He denied having any medical illness, trauma, or antecedent infection. On examination, his uvula was deviated to the right side and he had a decreased gag reflex. Laryngoscope revealed right vocal cord palsy with saliva accumulation in his piriform sinus (Figure). The laboratory data, which included complete blood count; C reactive protein level; erythrocyte sedimentation rate; and antibodies to Epstein-Barr viral capsid, cytomegalovirus, mumps virus, rubella virus, herpes simplex virus, my-

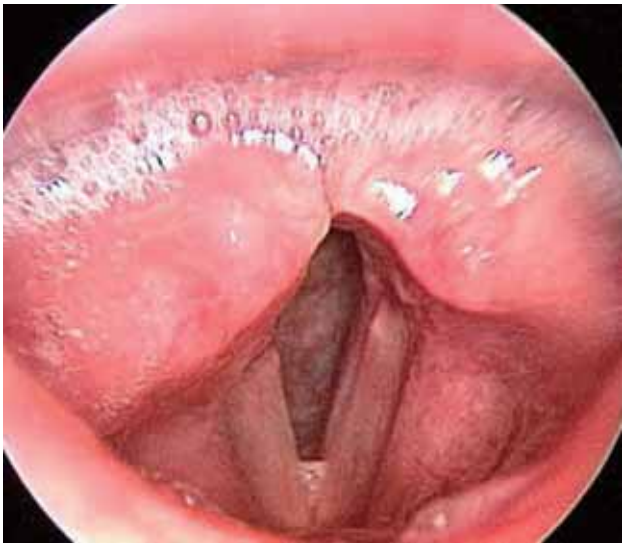

Fig. 1 Laryngoscopic photograph shows saliva pooling in right piriform sinus.

Received: March 9, 2010; Revised: April 15, 2010; Accepted: April 27, 2010
*Corresponding author: Chun-An Cheng, Department of Neurology, Tri-Service General Hospital, National Defense Medical Center, No. 325, Sec. 2, Cheng-gong Road, Taipei 114, Taiwan, Republic of China. Tel: 02-87927173; Fax: 02-87927174; E-mail: cca@ndmctsgh.edu.tw
Source of support: None declared.

coplasma, antinuclear, antineutrophil cytoplasmic, and anti-double stranded DNA, and rheumatoid factor, were within normal limits. Magnetic resonance imaging (MRI) showed no significant abnormal signal in the medulla. MR angiography revealed normal neck and intracranial vessels without plaque. MRI cranial nerve sequence showed well-delineated cranial nerves without abnormal enlargement.

Three days later, the patient complained of palpitation and dizziness after standing up. The resting heart rate in average increased from 68 to 76 beats/min and the range became more widely. Electrocardiogram demonstrated a normal sinus rhythm. Therefore, he received the head-up tilt test (HUT) with cardiovascular assessments including continuous beat-to-beat blood pressure and impedance cardiography. His heart rate was raised from 72 to 104 beats/min about 8 minutes after postural challenge. His systolic blood pressure increased from 118 to 128 mmHg and diastolic pressure from 75 to 88 mmHg. Transcranial Doppler sonography with a 2-kHz probe showed that cerebral blood flow velocity (CBFV) was reduced from 60 to 27 cm/s at the right middle cerebral artery. His cardiac output diminished from 5.3 to 4.7 L/min and total peripheral resistance on impedance cardiography increased from 1136 to 1724 dyn.s/cm^5. After recumbency, his heart rate returned to 65 beats/min and CBFV to 56 cm/s. The diagnosis of orthostatic tachycardia was established. Moreover, the beat-to-beat blood pressure recording with Valsalva maneuver with 40 mmHg for 15 seconds showed a normal phase II and overshoot IV response. Heart rate variability during deep breathing and the Valsalva ratio were below the normal limits compared with age-matched controls, which was compatible with parasympathetic impairment. His sympathetic skin response had a normal latency and amplitude. Then, he took propranolol 10 mg twice per day to inhibit sympathetic activity and piracetam 1200 mg per day to facilitate microcirculation. The nasal speech, hoarseness and orthostatic tachycardia remitted within three months. The laryngoscope showed adequate vocal cord motion without saliva accumulation in his piriform sinus. There was no relapse for 6 months.

DISCUSSION

Isolated unilateral vagal palsy is a rare disease that is mainly caused by trauma, surgery, toxins, infections, or medications. It often begins in the first or second decade of life, and 80% of victims are male.[2] The vagus nerves are originated form ambiguus nucleus in medulla, supplying the most of laryngeal and pharyngeal muscles. Regarding to the parasympathetic fiber, the right-sided vagus nerve can slow down the heart rate through sinoatrial node, and left-sided nerve can delay the atrioventricular conduction through atrioventricular node, respectively. Moreover, the afferent fiber of vagus nerve, carrying information from baroreceptor, cardiac receptors and chemoreceptors, are terminated in the nucleus of the solitary tract (NTS). Modality-selective NTS neurons are capable of communicating with caudal ventrolateral medulla (VLM), which can inhibit the sympathetic outflow from the rostral VLM to decrease peripheral vasomotor tone and cardiac excitability.[3] This patient had weakness of right-sided laryngeal and pharyngeal muscles and decreased parasympathetic activity with elevation of resting heart rate. Therefore, the pathological lesion should be located in the right-sided vagus nerve before the first branch to the pharynx. However, MRI and laboratory data cannot detect any structural lesion or remarkable inflammatory process.

The definition of postural orthostatic tachycardia syndrome (POTS) is an excessive increase in heart rate of 30 beats/min within the first 10 minutes of standing and associated with symptoms lasting more than 3 months.[4] The prevalence is estimated to be 170/100,000. Most patients develop orthostatic tachycardia in their teenage and then gradually improve after mid-twenties. Age of onset is most common between 15 and 50 years.[5] Females predominate over males by 4-6:1. Approximately one half of patients have antecedent viral or bacterial infection. The symptoms can be categorized into orthostatic (lightheadedness, palpitations, shortness of breath, and hyperhidrosis), autonomic (nausea, bloating, diarrhea, and abdominal pain), and diffuse associated (fatigue, sleep disturbance, migrainous headache, and myofascial pain). The patient, we presented, dose not fulfill the criteria of POTS owing to acute process and relatively short duration, but only orthostatic tachycardia. However, the pathophysiology in both of them should be similar.

The pathogenesis of orthostatic tachycardia is complex and multifactorial. One of the major hypotheses is that peripheral adrenergic denervation or restricted autonomic neuropathy results in decreased vascular sympathetic tone.[6] After the HUT, reduction of venous return causes raised heart rate for compensation of cardiac output. The lack of increase in peripheral vascular resistance as a consequence of adrenergic denervation results in decreased blood pressure and cerebral perfusion. A second hypothesis is that hypovolemia aggravates the decrement of preload after the HUT. Although the increas-

ing heart rate and vascular resistance can stabilize the blood pressure with only narrow pulse pressure, in the decompensated case, the blood pressure may eventually fall. A third hypothesis is that the hyperadrenergic state causes an overshoot of sympathetic tone and elevation of heart rate and blood pressure simultaneously.[7] Persistent tachycardia may decreases the ventricular diastolic filling time. When combined with increased peripheral vascular resistance, the reduction of stroke volume will develop. In our patient, increments in heart rate and blood pressure developed during postural challenge. Then, his end-diastolic volume index decreased from 70.3 to 46 ml/m^2 and total vascular resistance increased from 1136 to 1724 dyn.s/cm^5, which made stroke volume decrease from 86 to 49ml. Therefore, the hyperadrenergic state may be the most logical explanation for orthostatic tachycardia in this patient.

In addition, the patient developed orthostatic tachycarida fairly acutely following idiopathic vagal palsy, suggestive of high correlations between them. We reviewed the article about the pathogenesis of orthostatic tachycardia and POTS, and there is no report in parasympathetic dysfunction. As we know, the cardiovascular maintenance is based on the sympathetic and parasympathetic nervous systems, which act in a seesaw manner, with opposing actions on the target organ. Abolished parasympathetic activity may induce imbalance of homeostasis and disturbance of cardiovascular control. Therefore, the pathogenesis of orthostatic tachycardia may result from the parasympathetic dysfunction with a relative hyperadrenergic state.[8] To our knowledge, this is the first confirmed case of orthostatic tachycardia, caused by idiopathic vagal palsy with parasympathetic dysfunction.

Although orthostatic tachycardia is not a life-threatening disease, it may impair activities of daily living, similarly to other disabling conditions.[9] Patients presenting with vagal dysfunction should be warned about the possibility of developing orthostatic tachycardia. It is important to educate such patients to avoid aggravating factors such as dehydration, extreme heat, and vigorous exercise. Once orthostatic tachycardia is proven by the HUT, volume expansion with high salt intake and wearing an abdominal binder can alleviate the orthostatic symptom.[10]

In conclusion, patients with idiopathic vagal palsy should be carefully evaluated with regard to autonomic function. After confirming the diagnosis of orthostatic tachycardia, early health education and appropriate treatment can minimize functional impairment and improve quality of life.

ACKNOWLEDGEMENT

None declared.

REFERENCES

1. Stewart JM, Weldon A. Contrasting neurovascular findings in chronic orthostatic intolerance and neurocardiogenic syncope. Clinical Science. 2003;104:329-340
2. Alp H, Tan H, Altunkaynak S, Orbak Z. Idiopathic unilateral paralysis of the palate in childhood. Pediatr Neurol 2005;33:134-135.
3. Dampney RAL, Horiuchi J, Tagawa T, Fontes, MAP, Potts, PD, Polson, JW. Medullary and supramedullary mechanisms regulating sympathetic vasomotor tone. Acta Physiol Scand 2003;117:209-218.
4. Thieben MJ, Sandroni P, Sletten DM, Benrud-Larson LM, Fealey RD, Vernino S, Lennon VA, Shen WK, Low PA. Postural orthostatic tachycardia syndrome: the Mayo Clinic experience. Mayo Clin Proc 2007;82:308-313.
5. Low PA, Sandroni P, Joyner M, Shen WK. Postural tachycardia syndrome (POTS) J Cardiovasc Electrophysiol 2009;20:352-358.
6. Low PA. POTS. In: Low PA, Benarroch EE, editors. Clinical autonomic disorders. 3rd ed. Philadelphia: PA, Lippincott Williams & Wilkins; 2008. pp. 515-533.
7. Jordan J, Shannon JR, Diedrich A, Black BK, Robertson D. Increased sympathetic activation in idiopathic orthostatic intolerance: role of systemic adrenoreceptor sensitivity. Hypertension 2002;39:173-178.
8. Stewart JM. Autonomic nervous system dysfunction in adolescents with postural orthostatic tachycardia syndrome and chronic fatigue syndrome is characterized by attenuated vagal baroreflex and potentiated sympathetic vasomotion. Pediatr Res 2000;48:218-226.
9. Benrud-Larson LM, Dewar MS, Sandroni P, Rummans TA, Haythornthwaite JA, Low PA. Quality of life in patients with postural tachycardia syndrome. Mayo Clin Proc 2002;77: 531-537.
10. Grubb BP, Kanjwal Y, Kosinski DJ. The postural tachycardia syndrome: a concise guide to diagnosis and management. J Cardiovasc Electrophysiol 2006;17:108-112.