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Postural tachycardia syndrome associated with peripartum cardiomyopathy

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Abstract

Here we describe a 26-year-old woman who developed peripartum cardiomyopathy characterized by fatigue, poor exercise tolerance, and palpitations. Initial echocardiography showed global left ventricular dysfunction with an ejection fraction of 40%. Subsequently, the patient developed a resting tachycardia associated with symptomatic postural tachycardia. A diagnosis of Inappropriate Sinus Tachycardia was made based on holter monitoring studies with an average heart rate of 121 beat per minute (bpm). Standard autonomic evaluation confirmed the diagnosis of Postural Tachycardia Syndrome. The cardiomyopathy subsequently resolved (ejection fraction 60%, normal left ventricular function). However, both the postural tachycardia and sinus tachycardia persisted.

Keywords

postural tachycardia syndrome; pregnancy; peripartum cardiomyopathy

A rare complication from pregnancy is peripartum cardiomyopathy (PPCM). The underlying cause is unknown, but it is characterized by left ventricular dysfunction and symptoms of heart failure (Abboud et al., 2007). Here we present a case of a 26-year-old female who developed PPCM in the post partum period associated with the later development of Postural Tachycardia Syndrome (POTS) and Inappropriate Sinus Tachycardia (IST). To our knowledge this is the first such reported case on the subject.

A 26-year-old previously healthy female underwent a cesarean section at term (40 weeks) after developing a heart rate of 160 bpm. The remainder of the birth and subsequent brief hospital stay was uneventful. The child was healthy. The patient had no prior history of any cardiac disease. There was no family history of cardiac disease including early onset coronary artery disease, congenital heart disease, or sudden unexplained or cardiac death. The patient had been pregnant twice before with one healthy child and a spontaneous abortion at 8 weeks.

After delivery, the woman continued to have mild dyspnea and generalized fatigue with noted tachycardia (heart rate approximately 120 bpm). Approximately 3 weeks after delivery, the woman developed severe fatigue, activity intolerance, shortness of breath, and orthostasis consisting of lightheadedness and palpitations and presyncope. Echocardiography showed global left ventricular dysfunction with and ejection fraction of 40%. Extensive cardiac and endocrine work up (including negative ganglionic acetylcholine receptor and standard collagen

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vascular antibody panels) did not reveal any possible underlying cause and a diagnosis of PPCM was made. Sinus tachycardia (mean heart rate 121 bpm) was present on Holter monitoring studies without other arrhythmia types, consistent with the diagnosis of IST.

Standard autonomic testing confirmed the presence of postural tachycardia on head up tilt (Table 1, figure 1) associated with orthostatic symptoms confirming the diagnosis of POTS. Sudomotor and adrenergic function was intact including normal beat to beat blood pressure responses to valsalva maneuver. Cardiovagal function was diminished including a heart rate response to deep breathing range of 1.4 bpm (normal > 14 bpm; see figure 1 top panel) and valsalva ratio of 1.2 (normal > 1.4). Composite Autonomic Severity Scale (CASS) scores for each sub domain are given in Table 1.

The patient was treated with several beta-blockers with modest improvement of symptoms. PPCM had resolved approximately one year after onset with echocardiography studies being normal. The resting sinus tachycardia and postural tachycardia with orthostasis persisted. At this point (approximately one year after onset), the patient underwent invasive electrophysiology and radiofrequency ablation of the medial and lateral right atrium in an attempt to correct the IST. The procedure provided only transient relief and the tachycardia recurred 3 weeks after the procedure.

The patient continues to have difficulty with fatigue, activity intolerance, shortness of breath, and orthostasis. Treatment with a combination of increased fluids and salt intake, an incremental exercise regime, and medications (Nadolol 20 mg twice daily; Midodrine 5 mg three times daily) have improved her symptoms to the point that there is minimal impairment of activities of daily living. Repeat autonomic testing and heart rate measurements remain essentially unchanged 4 years after the initial diagnosis of POTS and IST.

POTS is defined as the development of orthostatic symptoms associated with a rise in standing heart rate of 30 bpm on head-up tilt and severe cases are associated with heart rate increments greater than 120 bpm (Low et al., 1994). Orthostatic symptoms most often include lightheadedness, palpitations, presyncope, and exacerbations with heat or activity (Thieben et al., 2007). The predominant age of onset is between the ages of 15 and 50 years, and there is a female predominance in this disorder (Low et al., 1994; Schondorf and Low, 1993; Thieben et al., 2007). Therefore, questions regarding the relationship between POTS and pregnancy are common in clinical practice. However, there is no published literature. In our laboratory we are currently undertaking a study on the relationship between POTS and pregnancy.

To our knowledge, based on an extensive literature search, this is the first case report of POTS associated with PPCM. There are prior reported associations of POTS with other cardiomyopathies (Khurana, 2008). However, evidence is limited to a handful of case reports. There is some commonality with regards to potential etiologies in that both POTS and PPCM have an association with viral infection. In POTS, 27.6% of patients report a prior preceding viral illness (Thieben et al., 2007). An underlying viral infection occurs approximately in 31–50% of patients with PPCM (Bultmann et al., 2005; Kühl et al., 2005). However, there was no reported viral illness in the case of our patient.

With regards to PPCM, several factors are important in relation to our case. Our patient had mild cardiac issues in comparison to many with PPCM. Prognostic factors in our case that predicted a good outcome included an ejection fraction greater than 30% as seen in our case (Ramaraj and Sorrell, 2009). It should also be noted that further pregnancy in women with a prior history of PPCM have a greater than 50% chance of relapse (Ramaraj and Sorrell, 2009).

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References

- Abboud J, Murad Y, Chen-Scarabelli C, Saravolatz L, Scarabelli TM. Peripartum cardiomyopathy: a comprehensive review. *Int J Cardiol* 2007;118:295–303. [PubMed: 17208320]
- Bultmann BD, Klingel K, Nabauer M, Wallwiener D, Kandolf R. High prevalence of viral genomes and inflammation in peripartum cardiomyopathy. *Am J Obstet Gynecol* 2005;193:363–365. [PubMed: 16098856]
- Khurana RK. Takotsubo cardiomyopathy in a patient with postural tachycardia syndrome. *Clin Auton Res* 2008;18:43–47. [PubMed: 17955329]
- Kühl U, Pauschinger M, Seeberg B, Lassner D, Noutsias M, Poller W, Schultheiss HP. Viral persistence in the myocardium is associated with progressive cardiac dysfunction. *Circulation* 2005;112:1965–1970. [PubMed: 16172268]
- Low PA, Opfer-Gehrking TL, Textor SC, Schondorf R, Suarez GA, Fealey RD, Camilleri M. Comparison of the postural tachycardia syndrome (POTS) with orthostatic hypotension due to autonomic failure. *J Auton Nerv Syst* 1994;50:181–188. [PubMed: 7884158]
- Ramaraj R, Sorrell VL. Peripartum cardiomyopathy: Causes, diagnosis, and treatment. *Cleve Clin J Med* 2009;76:289–296. [PubMed: 19414544]
- Schondorf R, Low PA. Idiopathic postural orthostatic tachycardia syndrome: an attenuated form of acute pandysautonomia? *Neurology* 1993;43:132–137. [PubMed: 8423877]
- 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. [PubMed: 17352367]

Figure 1.

Autonomic function tests. Heart rate response to deep breathing was attenuated (A). Heart rate increase was excessive in response to head up tilt (B). In addition, beat-to-beat blood pressures during head up tilt showed significant variability compatible with orthostatic intolerance (B). The respiratory rate is indicated by the blue line. The heart rate is indicated by the green line. The beat-to-beat blood pressure is indicated by the red line. The y axis represents blood pressure (left) and heart rate (right). Time in minutes is expressed on the x axis. The black line indicates duration of head up tilt (B).

Table 1

Autonomic and cardiac test results

CASS total	2			
CASS sudomotor	0			
CASS cardiovagal	2			
CASS adrenergic	0			
Heart rate supine (bpm)	132*			
Heart rate standing 5 min. (bpm)	165*			
Heart rate_Δ	33*			
supine SBP (mmHg)	116			
supine DBP (mmHg)	70			
standing SBP (mmHg) 5 min.	120			
standing DBP (mmHg) 5 min.	78			
Δ systolic	4			
Δ diastolic	8			
NE supine	327			
NE standing	715			
NE_Δ	388			
24 hour urine volume (mL)	1298			
24 hour urine Na ⁺ (mmol/L)	206			
		<u>3 weeks</u> [†]	<u>1 year</u> [†]	<u>2 years</u> [†]
Left ventricular function	global dyskinesia	normal	normal	
ejection fraction	40%	60%	60%	

* at diagnosis;

[†] post partum;

CASS, Composite Autonomic Severity Score; SBP, systolic blood pressure; DBP, diastolic blood pressure; NE, norepinephrine