CLINICAL VIGNETTE

Postural Tachycardia Syndrome: A Case Review

Suruchi Kelly, M.D.

Case Presentation

A 22-year-old female presents with a 5-year history of dizziness upon standing, generalized weakness, headaches, nausea sometimes accompanied by vomiting, and tachycardia. She has been seeing a gastroenterologist and full evaluation of her GI tract reveals no structural abnormalities. She feels like fainting as long as she is upright, and this feeling improves when she lays down. She notes that her pulse increases from 80 to 140 bpm when she goes from a sitting to a standing position. She has been diagnosed with Raynaud's phenomenon as her fingers can turn cold and blue. She has difficulty concentrating and reports tinnitus. Her menstrual cycles are almost absent as she has an IUD.

Vital signs on exam include sitting blood pressure 89/65 and pulse 104, height 5 ft 10 inches, weight 126 lbs, and BMI 18.

She appears euvolemic by exam. Lungs clear. Heart sounds regular rate and rhythm without murmurs. Abdomen with normoactive bowel sounds, no tenderness or distention, and no organomegaly. Extremities with some mild cyanosis. Neurologically, her cranial nerve 2-12 functions are intact. Her Romberg is negative. Her tandem gait intact.

Her ECG showed NSR rate of 61 and incomplete bundle branch block. Her stress ECHO was normal, with no inducible arrhythmias upon exercise. Her CBC and TSH were normal and her Cosyntropin stimulation test showed appropriate increases in cortisol levels.

She was seen by Neurology and diagnosed with postural tachycardia syndrome.

Discussion

Postural tachycardia syndrome (POTS) usually presents between ages 13-50 and is more likely to occur in females than males (80-85%).[1] Etiology is thought to be multifactorial. It manifests with cardiac symptoms such as dizziness, palpitations, chest pain, dyspnea or fatigue, as well as non-cardiac symptoms like blurry vision upon standing, headache, mental clouding, and anxiety. GI symptoms are also common and include nausea, cramps, early satiety, bloating, and constipation. Acrocyanosis and extremity edema can be visible upon patient standing. Forty percent of patients will experience syncope.[1,2]

Certain criteria need to be met in order to make the diagnosis.[1,3]

1) Heart rate increase over 30 bpm from supine to standing over a 10 minute period.
2) Symptoms worsen with standing and improve with a recumbent position.
3) Symptoms have been ongoing for over 6 months.
4) Absence of other overt causes of orthostatic symptoms or tachycardia (dehydration, medications, anemia, hypothyroid, etc.).

Normal physiology dictates that when a patient assumes an upright posture, there is a shift of 500 ml of blood volume to the dependent areas, such as abdomen and legs. This shift decreases venous return and cardiac output and eventually blood pressure. This "unloads" baroreceptors and triggers a reflex sympathetic activation resulting in elevation of heart rate by 10-20 bpm and systemic vasoconstriction. In comparison, the POTS patients have an exaggerated response to the drop in blood volume in terms of heart rate and sympathetic tone.[1]

POTS and vasovagal presyncope can look very similar but have some distinct differences. Although vasovagal syncope can present at any age, it tends to manifest in the second or third decade of life. It is less common in women than POTS. It tends to occur after prolonged sitting or standing, whereas POTS comes on immediately with sitting or standing. Vasovagal patients more commonly faint and only feel orthostatic at the time of the faint. POTS patients can feel orthostatic throughout the day. On tilt table test, the vasovagal patient exhibits a drop in blood pressure and pulse whereas POTS patients become tachycardia and may have unchanged blood pressure.[3,4]

Many proposed etiologies exist for POTS. Neuropathic POTS occurs due to denervation of the sympathetic nerves in the lower legs. Hypovolemic POTS may be due to abnormalities in the renin-angiotensin-aldosterone system leading to low aldosterone. Hyperadrenergic POTS is caused by elevated norepinephrine serum levels usually >600 pg/ml while standing. These patients may have high blood pressures upon

standing. Other POTS patients have a mast cell activation disorder where their mast cells degranulate without any obvious trigger, resulting in episodic flushing and increased urine methylhistamine levels. Another type of POTS can be genetic, due to a mutation in the norepinephrine transporter, resulting in decreased clearance of synaptic norepinephrine.[1,3] It is notable that some psychiatric medications can lead to inhibition of the same norepinephrine reuptake transporter and mimic the POTS effect. There has been association with acetylcholine receptor antibodies in 10% of POTS patients, suggesting an autoimmune association.[2]

Diagnosing POTS should include an orthostatic challenge either with tilt table or upright posture. ECG should confirm sinus tachycardia, not other arrhythmia. A Holter monitor could be considered. ECHO should show no structural abnormalities. Measurement of the norepinephrine level while standing should be taken. CBC, electrolytes, thyroid panel, celiac panel, B12, and iron are recommended to rule out other causes of orthostasis, tachycardia, and GI symptoms.

Treatment includes consumption of 8-10 glasses of water daily and increasing sodium intake to 8-10 gms daily. If this is not possible through diet, addition of NaCl tablets can be used. Use of compression hose, preferably waist-high, can enhance venous return. Exercise has been routinely recommended but may not be tolerated by all POTS patients. It has been shown to decrease the orthostatic tachycardia.[3]

So far FDA has not approved any medications for POTS, but certain medications have been proven useful when used "off-label." In patients suspected to have hypovolemia-induced POTS, fludrocortisone is beneficial at a dose of 0.1-0.4 mg daily. Side effects can include supine hypertension, fluid retention, and hypokalemia.[3]

In patients with neuropathic POTS, midodrine, an adrenoreceptor agonist, can help to vasoconstrict vessels. Side effects include goose bumps, scalp tingling, or headaches.[1] Propranolol can help reduce the tachycardia but should not be used in patients where the tachycardia is a compensatory response to low stroke volume. It also can worsen symptoms in a patient with mast cell activation disorder. Clonidine 0.1-0.2 mg BID to TID can stabilize blood pressure and heart rate in patients with high sympathetic nervous system activity. Side effects can include drowsiness and fatigue and trouble concentrating. Caution should be used in prescribing any drosperinone-containing oral contraceptive, a spironolactone-analogue, which can lead to low aldosterone and low blood volume.[1]

Conclusion

Our patient was started on Fludrocortisone, increased fluids, salt tablets, and increased exercise. She felt symptoms had improved significantly but moved out of state and was lost to subsequent follow-up.

REFERENCES

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