Postural Orthostatic Tachycardia Syndrome

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Case Overview

POTS is a little known condition affecting the autonomic nervous system, and 80-85% of those affected are female, with usual onset at puberty.

Presenting Complaint

LM is a 17-year old adolescent female in Pennsylvania whose parents sought medical care at age of 12 after a week of tiredness and headaches which persisted and did not respond to home interventions. LM first displayed symptoms at age 12, and these consisted of extreme fatigue, heart palpitations, sleeping 15 to 17 hours a day, headaches, dizziness, and inability to attend school without feeling sleepy.

Patient Profile

- Name: LM
- Age: 17
- Sex: Female

Social Status: Single adolescent living at home with parents and an older sister.

Occupation/Lifestyle: Student-12th grade, not working currently. The main limitation is not being able to stand up for long periods of time. At school she has a plan that allows her to start the school day later and modified PE class. Additionally, she has water and salty snacks available at all times as well as extra time to complete work when absent from school for more than a week.

Medical History: Patient has been diagnosed with: POTS (Postural Orthostatic Tachycardia Syndrome), IBS (Irritable Bowel Syndrome), GERD (Gastro-Esophageal Reflux Disease), Dysmenorrhea, Hypersomnia, and Overactive Bladder. Patient suffers from fatigue, heat intolerance, tachycardia, low blood pressure, low blood volume, blood pooling, dizziness/light-headedness, abdominal pain, and flushing.

Family History: No history of neurologic or hypotensive or autonomic disorders.

Interventions: Florinef, Midodrine, Seasonique, Amitriptyline, Ritalin, Dicyclomine, and Omeprazole.

Non-pharmaceutical treatments; melatonin, physical therapy, yoga, acupuncture, vegan diet, weight-lifting, salt tablets, bed risers, increased salt and water intake, compression stockings, probiotics, and recumbent bike.

Assessment

Patient has been evaluated by pediatrics, cardiology, neurology, gynecology, nutritionist, physical therapists, rheumatology, urology, pediatric gastroenterology, and adolescent medicine.

Blood work: Mononucleosis screen, Lyme Disease screen, Complete Blood Count, Comprehensive Metabolic Screen, Phosphorus, Ferritin, Epstein-Barr Antibody, RBC SED Rate, Insulin, Glucose, Blood Ventilation/Perfusion Ratio, Thyroid, Thyroxine.

Other Testing: Echocardiogram, Head CT scan, Poor Man's tilt-table, Sleep study, Transthoracic Echogram, Tilt-table test, QSART (Sweat test), Endoscopy, Esophageal pH Probe test, Renal ultrasound scan, and Urinalysis.

Pediatrics initially diagnosed a post viral syndrome but blood work did not confirm so other tests were required and patient was referred to a cadre of specialists.

- Cardiology specialist diagnosed POTS.
- Neurologist made a secondary diagnosis of mild Neurogenic orthostatic hypotension with neurogenic POTS.
- Pediatric gastroenterologisy diagnosed with IBS and GERD.

Diagnosis

Postural Orthostatic Tachycardia Syndrome (POTS). A little known, yet fairly common Dysautonomia Syndrome. (describing various medical conditions causing malfunction of the autonomic nervous system) For more information, see:
http://dysautonomiainternational.org/welcome.php

Evaluation/Treatment

Initially, two different pediatricians were not able to determine a diagnosis. One of them suspected an anxiety disorder or school aversion. After several visits, and numerous blood tests, patient was referred to a cardiologist and a rheumatologist. The cardiologist was the first one to evaluate for, and diagnose POTS. Patient's fatigue and change in sleep cycle were not addressed by this diagnosis. A rheumatologist confirmed the diagnosis and referred patient to a sleep clinic. A sleep study result was normal.

Patient was started on fludrocortisone, increased fluids, and salt. The fludrocortisone helped initially, but improvement did not persist.

A second Cardiologist started midrodine, an alpha receptor agonist.

Due to dysmenorrhea, gynecology prescribed Seasonique, (Levonorgestrel/Ethinyl Estradiol – extended cycle). The patient states it is her "magic" pill. Very effective for the dysmenorrhea. Patient was unable to attend school for nearly a year and was bed ridden. After ~ 6 months patient saw a neurologist in Florida. The neurologist diagnosed Mild Neurogenic orthostatic hypotension with neurogenic POTS. Neurologist suggested compression stockings, raised head

of bed, salt tablets, and resistance training for deconditioning. Ultimately, she saw a cardiologist in Ohio who specializes in treating POTS. He prescribed methylphenidate which only helped the symptoms for a week. She experienced side effects of appetite suppression and abdominal pain.

Some non-pharmacologic treatments were recommended: physical therapy, salt tablets. Physical therapy was difficult due to deconditioning and low blood pressure which limited this modality.

Because of persistence of abdominal pain, referral was made to gastroenterology. Additional tests: Endoscopy and Ph probe were normal. Patient was prescribed dicyclomine (Bentyl), and omeprazole. The omeprazole didn't give any relief of GI symptoms, and initially dicyclomine at 10 mg. was no help.

Adolescent Medicine Referral suggested amitriptyline (Elavil), 20 mg. for the abdominal pain, but not much improvement. When this was increased to a higher dose (35 mg.), the abdominal pain subsided somewhat. An additional increase of the dose to 50 mg. has helped remarkably. Again, the patient dubs these her "life savers."

Discussion

The presentation and course of this adolescent female with POTS is likely typical, as it is a complex disorder with symptoms which require expertise from many disciplines of medicine, as many syndromes do. This girl presented with multiple symptoms, and faced the skepticism that many women experience when their symptoms don't fit a particular known diagnostic pattern. As many women have experienced, when symptoms aren't standard, a diagnosis of anxiety or other psychologic spectrum is often assumed, and, indeed, counseling was recommended for this patient, but wasn't particularly helpful.

Since her symptoms spanned gyn, GI, neurology, cardiology, and urology, it was hard for the parents to decide where to start, and they needed to persist in researching symptoms, consulting support groups, and finding the proper diagnostician. And even then, several treatment options needed to be tried before finding any that were helpful. POTS often presents in adolescence, and is much more prevalent in girls (5:1, female:male) and since much of the diagnostic testing and symptomatology lies within the purview of cardiology, the consultation in that field turned out to be the most helpful. In fact, the parents were able to find an expert cardiologist in Ohio who specializes in these cases. Unfortunately, the etiology is still largely unknown, but it seems to be associated with an immune-mediated neuropathy, with studies showing autoantibodies to alpha3 acetyl choline receptors of the peripheral autonomic ganglia. This would likely explain the preponderance of diagnosis in females, as many autoimmune diseases seem to be more common in women.

Key Words: Case Study, postural orthostatic tachycardia syndrome, POTS, Sex and Gender Medicine