A Difficult Diagnosis: POT Syndrome Presenting with Severe Abdominal Symptoms

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Abstract
Postural orthostatic tachycardia syndrome (POTS) is a disorder of the autonomic nervous system. Classically, it is characterized by symptoms of postural hypotension and tachycardia in the erect position, however a wide range of other symptoms are recognized. It can be difficult to diagnose with patients having multiple negative investigations and eventually being misdiagnosed as having psychiatric disorders or chronic fatigue syndrome (2). It can affect any age or sex but usually tends to occur in those aged between 15 to 50 years, with a female bias (4). A case is presented where the patient had a significant number of investigations for ongoing abdominal symptoms.

CASE REPORT
A 44 year old male was referred to the Cardiology Department for an in-patient opinion by the Gastroenterology team. He was admitted electively for investigation of a number of symptoms including nausea, epigastric pain, dyspepsia, abdominal bloating, cold extremities, fatigue, palpitations, postural instability, anxiety, insomnia and breathlessness. His symptoms had initially started in 2000, and had been getting worse with time, such that despite being an athlete with a rigorous personal exercise program, he was now confined to home and on long term sick leave.

Prior to admission, the patient had already had surgical, cardiology, psychiatry, rheumatology, gastroenterology, neurology and pulmonology consults. Interventions in the early and ongoing stages of the patients' disorder had focused on the abdominal symptoms as these were considered to be the most severe. The patient had surgical intervention with laparoscopic fundoplication, after being found to have a moderately sized hiatus hernia with mild gastric reflux, and failing high dose therapy with proton pump inhibitors. Unfortunately, this intervention had no effect on the patients' quality of life, and symptoms deteriorated further. For this admission, the Gastroenterology team arranged endoscopies, nuclear gastric emptying studies, porphyria screen, MRI scan of brain and spinal cord, pulmonary function tests, and CT abdomen. All tests were normal.

During his stay the patient developed an episode of chest tightness, and tachycardia. An myocardial infarction screen was negative and an ECG confirmed sinus tachycardia. Examination of the patient was normal, and the previous normal investigations were noted. The only interesting finding was persistent tachycardia when the patient stood up (from 130-140bpm) with a slight drop in blood pressure. It was this and the extensive list of normal and negative investigations which raised the diagnosis of POTS. It was also noted that at the beginning of a previous exercise treadmill test the patient developed tachycardia, and low blood pressure which was attributed to anxiety. Previous 24 hour ECGs also revealed runs of sinus tachycardia, which co-related with symptoms and which had been attributed towards anxiety.

The patient was informed of the diagnosis and given lifestyle advice as well as a therapeutic trial of bisoprolol. He attends clinic for regular review and is currently doing well with a significant reduction in symptoms.

DISCUSSION
This case illustrates the difficulty in diagnosing POTS, especially when numerous specialties have been involved, and no clear answer is obtained from investigations. The significance of postural tachycardia in this case was overlooked, thereby delaying diagnosis. The patients' marked abdominal symptoms played a role in making diagnosis difficult. Unfortunately, POTS can present with a variety of symptoms as this case demonstrates (1,5).
To make the diagnosis of POTS there has to be a heart rate increment of at least 30bpm within 5 minutes of standing, with heart rate over 120bpm and orthostatic symptoms. However, variants are recognized where blood pressure may be high or even normal, although postural tachycardia is usually a consistent finding (S10). Tilt table testing is useful to confirm these findings in equivocal cases. More specialized tests include measuring red cell mass and venous pooling in the calf muscles with isotopes, as well as using isoproterenol (beta receptor agonist) during tilt testing to demonstrate beta receptor hypersensitivity and stimulate symptoms. Again baseline tests e.g. ECG, ECHO, routine bloods etc are important to rule out primary cardiac and neurological diseases (S10).

POTS is poorly understood, with several mechanisms proposed to explain the phenomenon. This includes beta receptor supersensitivity, erythropoietin deficiency, reduced red cell mass and low circulating blood volumes due to defective peripheral vasoconstriction leading to pooling of blood and relative hypovolemia, as well as genetic factors in some families (S10). Additionally, in some patients the condition has occurred following viral infections, suggesting a possible immune mediated response (S10). Occasionally, POTS can be secondary to other disorders such as diabetes, amyloidosis, SLE or Multiple System Atrophy (S10).

Treatment involves patient education, lifestyle changes such as increasing fluid and salt intake, physiotherapy and graduated training, along with drug therapies (addressing the proposed mechanisms of POTS). Various drugs have been used to increase circulating volume such as fludrocortisone and erythropoietin. Yet other medications have been used because of their centrally acting and vasoconstrictive responses such as clonidine, midodrine, venlafaxine and methydopa. Some patients may benefit from beta blockers if they are thought to be beta receptor super sensitive (S10).

CONCLUSION

This case highlights that POTS is a difficult diagnosis to make, & a high degree of clinical suspicion is needed to make the diagnosis, especially when gastrointestinal symptoms prevail. As illustrated POTS can dramatically affect quality of life, causing significant disability if not picked up and treated (S10).

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References

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