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Case Report

POTS Treatment Complicated by Clots, Pulmonary Embolism and Endocarditis

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Abstract

POTS (Postural Orthostatic Tachycardia Syndrome) is defined as an increase in heart rate of greater than or equal to 30 beats/minute within the initial 10 minutes of standing or Head-Up Tilt (HUT) in the absence of orthostatic hypotension. Patients with POTS can exhibit hypovolemia with treatments including IV fluid resuscitation, salt tablets and medication for heart rate control. Fluid resuscitation can increase risk for clotting, endocarditis, and pulmonary embolism. This retrospective case review will discuss three cases of patients with POTS who had complications with access lines leading to clots, infections, endocarditis, and pulmonary embolism.

Keywords: Postural orthostatic tachycardia syndrome; Infection; Pulmonary embolism; Autonomic dysfunction; Infective endocarditis

Introduction

The diagnosis of POTS is made by a history of orthostatic intolerance with or without systemic symptoms, and a correlation of symptoms with a sustained increase in upright heart rate by at least 30 beats/minute (or 40 beats/minute for patients under 20 years of age) within initial 10 minutes of standing or Head-Up Tilt (HUT) without orthostatic hypotension. POTS predominantly affects premenopausal females (5:1 ratio) between 13 and 50 years of age, manifesting with symptoms of fatigue, headache, palpitations, sleep disturbances, nausea, or bloating [1]. Patients may develop POTS after a viral illness, infection, pregnancy, or trauma of the head. Chronic autoimmune diseases like Sjogren's and Celiac are often associated with a POTS diagnosis. Among genetic disorders, POTS has been linked to joint hypermobility disorders including but not limited to Ehlers-Danlos Syndrome (EDS). Notably the presence of EDS is higher in patient with POTS than the general population [2].

The pathophysiology behind POTS is largely unknown. A multitude of mechanisms have been proposed including disproportionate sympathoexcitation, volume depletion, autoimmune dysfunction, and cardiac and physical deconditioning [1]. Patients with POTS are unable to coordinate balancing blood vessel constriction with heart rate response, leading to the lightheadedness, fainting, and rapid increase in heartbeat when standing [3]. There are multiple forms of POTS, including neuropathic, hyperadrenergic, hypovolemic, and secondary POTS.

Treatment of POTS has been directed at symptom management, responding to a combination of diet, medications, physical therapy and other treatments like cardiovascular reconditioning, horizontal exercise (rowing, swimming), use of compression garments and volume expansion via increased salt and fluid intake [4]. There is no universal pharmacological treatment. Pharmacological treatments include fludrocortisone, pyridostigmine, midodrine and beta blockers [5]. Fluid resuscitation may be used in extreme cases where GI fluid absorption is impaired. Line infections, sepsis, and Infective Endocarditis (IE) may be seen. Classically, IE is an infection of the native or prosthetic heart valves, but with an increasing number of intracardiac devices and central lines, there is an increase in the incidence of the intracardiac abscess (endocardial abscess) and catheter-related infections. POTS patients may also have clots in their lines or in other areas of their bodies, including Pulmonary Embolism (PE). This is a disruption to the flow of blood in the pulmonary artery or its branches by a thrombus that originated somewhere else [6]. In this retrospective case series, we present three cases of patients with POTS who had complications with access lines that lead to clots, endocarditis and pulmonary embolism.

Case Presentation

Case 1

Patient 1 is a 34-year-old that started exhibiting symptoms of syncope at age 27. They continued to experience these symptoms with associated tachycardia which led to a hospitalization and a diagnosis of POTS. Notable medical history included epilepsy and gastroparesis. Additional neurological testing revealed a small fiber neuropathy. The patient did not tolerate selective beta blockers and was initiated on pyridostigmine. Due to recurrent hypovolemia, the patient was started by an outside provider on maintenance fluids. This was initially one liter of normal saline once a week and it increased over time to several times a week. Due to access issues to deliver fluids, a central line was placed. She developed gastroparesis and required TPN administration. There were three episodes of sepsis associated with the central line. These sepsis events resulted in the line being pulled and IV antibiotic initiation during hospitalizations. When fluids were stopped during sepsis, the patient had increase in episodes of syncope. One episode resulted in a fall with a leg fracture.

Case 2

Patient 2 is a 47-year-old female that was diagnosed with a variant

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form of Ehlers Danlos type with brittle corneal changes (ZnF469). Initial presenting symptoms included nausea, diaphoresis even with slight exertion, and tachycardia. Autonomic testing interpretation: Abnormal study. The results are consistent with: Sudomotor failure which can be seen in mild small fiber neuropathy since the sudomotor testing was abnormal, but the skin biopsy was normal. Normal adrenergic and cardiovagal functions were noted. Orthostatic cerebral blood flow was associated with hypocapnia without orthostatic hypotension. Hypocapnic hyperventilation was noted during the tilt. She was hospitalized multiple times for syncope at the onset of the POTS diagnosis and treated with IV fluids with improvement. In the first year of her POTS diagnosis the patient developed her first line infection and was treated with antibiotics. She subsequently required a port for total parenteral nutrition and IV fluids. Subsequently, the access site had multiple issues with clots, requiring increasing size of the catheter for access. One year following the POTS diagnosis the patient was admitted for bilateral pulmonary emboli and was initiated on enoxaparin. The admission was followed by a port infection (blood cultures with Staph Epidermidis), and a line infection that was treated with antibiotics. In the second year after her POTS diagnosis, a V/Q scan displayed bilateral pulmonary emboli, with clots in bilateral lower branches of lungs. She was started on fondaparinux. In the ensuing years she developed one line infection and four port infections. Patient 2 was eventually placed on a powerport which is an implantable catheter used to deliver fluids and medication.

Case 3

Patient 3 is a 30-year-old female who began exhibiting syncopal episodes associated with general malaise at 12 years of age. It took 7 years to establish the diagnosis of POTS through the results of tilt table testing. The autonomic function testing demonstrated neuropathy affecting sympathetic adrenergic fibers; the patient developed presyncope 6 minutes after upright tilt testing with a significant drop in blood pressure and blood flow. Pertinent patient medical history included Ehlers Danlos and mitochondrial disorder. Over the course of her diagnosis, the patient had fifteen separate IV lines placed which including PICC lines, tunneled PICC lines in the chest, and ports for TPN. There were four major infections where lines or ports were removed and she received antibiotics. Most lines were placed for six months to a year. The first major complication was a septic infection with a clot in her line resulting in treatment with antibiotics and apixaban. Subsequently she suffered from severe septic endocarditis and pneumonia. She required a porcine tricuspid valve placement due to the septic endocarditis. Additionally, she developed a clot in an IJ line. Recurrent use of indwelling lines in this patient with POTS resulted in complications of clots, infections, and endocarditis requiring a valve replacement.

Discussion

This case series discusses three patients with diagnosed postural orthostatic tachycardia syndrome who had treatment complications including infections, line clots, pulmonary emboli, and endocarditis (Table 1). POTS proposed mechanisms include disproportionate sympathoexcitation, volume depletion, autoimmune dysfunction, and cardiac and physical deconditioning [1]. The mainstays of treatment for patients that have POTS are supportive care including salt tablets and electrolytes, adequate hydration, and heart rate stabilization. The mechanism in which infections can arise from intravenous access includes the migration of microbes down a catheter tract, inadequate asepsis from care providers, and improper maintenance of dressings [7]. It is well known that catheter dwell time is one of the major risk factors for PVC infection, but the removal of the IVC does not reduce infection risk, thus simple exposure increases infection risk overall [7]. Intravenous catheters are known to cause endothelial trauma and it has been discovered that POTS patients are more hypercoagulable [8]. The pathophysiology of POTS is also commonly linked to other syndromes and diseases requiring the need for nutrition support as patients have an increased risk of having concomitant gastroparesis and autonomic dysfunction. Optimization of gut absorption, as feasible, may mitigate the need for recurrent IV fluids. In those who are unable to achieve euvolemia due to limited gut absorption, care must be taken in placing and monitoring indwelling lines. Diligent monitoring and management may reduce risk of infection, clots, and emboli, but the danger remains.

1111: Total number of episodes of infections, clots, and endocarditis in each patient.

Patient	Infections	Clots/PE	Endocarditis
1	6	0	0
2	8	2	0
3	6	1	1

Statement of Ethics

This study protocol was reviewed and determined to be exempt by the St. Elizabeth's Medical Center IRB, on February 15th, 2022. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

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Author Contributions

Conceptualization: A.H; Writing of manuscript: L.M; M.C. Data Collection: L.M, A.H; Formal Analysis: L.M; M.C.; Review and Editing: A.H.

Data Availability

Details were found in the patient's medical record and on no other publicly accessible website. All data generated or analyzed during the study are included in the article. Further enquiries can be directed to the corresponding author.

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