E17. EASILY MISSED: RESISTANT POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME IN A HYPERMOBILE YOUNG MAN

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Background: Postural orthostatic tachycardia syndrome (POTS) is characterized by the onset of orthostatic symptoms with a rise in heart rate of ≥30 beats/min within 10 min of standing or an absolute heart rate of >120/min following tilt-table testing (gold standard investigation) at an angle of about 60–70 degrees to the horizontal. It is highly associated with joint hypermobility syndrome (JHS) or Ehlers-Danlos syndrome hypermobility type as a form of autonomic dysfunction. Individuals affected are mainly aged between 15 and 50 years and predominantly female. Often under-recognized, POTS may mimic other conditions such as anxiety or neurocardiogenic syncope. Management of POTS is difficult, as there is limited evidence of the potential therapeutic benefits from both pharmacological and non-pharmacological approaches.

Methods: A 23-year-old man presented to the rheumatology clinic with hypermobile joints and a 6-month history of recurrent palpitations, light-headedness and lower limb weakness upon standing without any associated syncope, which improves on recumbence. However, his functional ability has been impaired. Previous encounters of similar episodes were diagnosed as chronic fatigue syndrome. His medical history includes childhood blackouts, depression and FM for which appropriate medications are taken.

Results: On clinical examination, his heart rate increased from 80/min [blood pressure (BP) 147/97 mmHg] when supine to 146/min (BP 142/87 mmHg) when standing for 10 min. He was also noted to have hypermobile joints in all limbs and cervical spine, fragile skin, bilateral pes planus and thoracic scoliosis, all consistent with JHS. His blood work-up (full blood count, urea and electrolytes, RF, ANA, CRP, liver function tests, thyroid function tests) was unremarkable. Transthoracic ECG revealed laxity of mitral valve and surrounding ligaments. Furthermore, tilt-table testing at 60 degrees displayed a significant heart rate increment from lying (53/min, BP 113/68 mmHg) to standing (140/min, BP 124/86 mmHg). As such, JHS-associated POTS was diagnosed. He was started on a trial of midodrine 2.5 mg and sodium chloride 600 mg for 3 months but these worsened his symptoms. Thereafter, fluoxetine 20 mg and labetalol 50 mg were tried but to no avail. Hence, he was managed conservatively, aimed at symptom control. Occupational and physiotherapy support was also sought with muscle-strengthening exercises. Advice was also given about optimizing fluid and salt intake.

Conclusion: A high index of suspicion of POTS should be made in hypermobile patients presenting with orthostatic intolerance. Diagnosis can be complicated due to overlapping features with other conditions. It is imperative that POTS be recognized and treated promptly as symptoms can be debilitating. Thus far, no single therapy has been proven effective. Non-pharmacological measures should first be considered in otherwise fit and healthy individuals. In more severe symptoms or associated vasovagal syncope, pharmacological (e.g. fludrocortisone, selective serotonin re-uptake inhibitors, pyridostigmine) intervention should be considered. A multidisciplinary holistic care must be employed in treating non-autonomic features (e.g. fatigue, musculoskeletal damage) and tailored to patients’ needs.

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