Unusual presentation of more common disease/injury

Unusual cause of acquired brain injury

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A 37-year-old male was admitted with history of syncope and fall leading to right temporal bone fracture with extradural haemorrhage. He underwent craniotomy and clot evacuation. Postoperatively during inpatient rehabilitation, he was progressing well with his physical and cognitive rehabilitation, when he had resting tachycardia of between 90 and 100 per minute. Investigations to rule out cardio-pulmonary and endocrine causes of tachycardia were normal. He was further investigated for suspected postural orthostatic tachycardia syndrome (POTS). A tilt table test was done which revealed rise in baseline heart rate from 90/min to 130/min on 70 with no postural drop in blood pressure. Subsequently plasma norepinephrine level in erect posture was analysed which was 721 pg/ml. A working diagnosis of POTS was made. Initially, after an increase in water intake by 1.5–2 l/day, his baseline and postural tachycardia reverted back to 76/min. He is being followed up.

BACKGROUND

Postural orthostatic tachycardia syndrome (POTS) is commonly recognised condition in specialised units where

multidisciplinary care is done for similar conditions. A combined effort of cardiologist, neurologist, endocrinolo-

Figure 1 CT brain on admission: right extradural haemorrhage.

Figure 2 CT brain on admission: right extradural haemorrhage with midline shift.

gist and physiologist helps to identify this condition and manage easily.

Despite this, among common medical practice this is quite an under diagnosed medical problem.

We wish to highlight that POTS although common but can be missed due to lack of awareness leading to increased suffering for the patient and multiple referrals with non-specific presentations.

POTS is easy to diagnose and manage with simple measures.

CASE PRESENTATION

A 37-year-old male was admitted with history of syncope (what appeared to be micturition syncope) and fall leading to head injury.

INVESTIGATIONS

An urgent CT scan of brain revealed; right temporal bone fracture with extradural haemorrhage and mass effect (figures 1 and 2).

He underwent emergency craniotomy and clot evacuation. Postoperatively he was transferred for inpatient rehabilitation.

While he was progressing well with his physical and cognitive rehabilitation, he was noticed to have resting tachycardia of between 90 and 100 per minute.

Investigations to rule out cardio-pulmonary and endocrine causes including: full blood count, renal function tests, adrenal, thyroid function, infection screen, echocardiography, 24 h Holter monitor, screen for pulmonary embolism were normal.

As his ECG showed right bundle branch block pattern, due to his young age and history of syncope: Brugada syndrome was ruled out with negative electrophysiological test (Flecainide challenge test).

His medical history was negative for: diabetes, smoking, alcohol intake and any other endocrine or cardiac problems.

He was not on medications including vasodilators, anti-depressants and anxiolytics.

He had history of vague tiredness, easy fatigability for 2 years.

He was further investigated on our unit for suspected POTS. A tilt table test was done which revealed rise in baseline heart rate from 90/min to 130/min on 70 with no postural drop in blood pressure (figure 3).

Subsequently plasma norepinephrine level in erect posture was analysed which was 721 pg/ml.

A working diagnosis of POTS was made.

Figure 3 Tilt table: graph of blood pressure, heart rate against degree of tilt.

TREATMENT

Initially, he was advised to increase his water intake by 1.5–2 l/day. His baseline and postural tachycardia responded and reverted back to 76/min with no BP changes.

With no further symptoms of tiredness and fatiguability, he was given careful counselling as well as explanation of the diagnosis and further management for future.

OUTCOME AND FOLLOW-UP

He returned to full time work with no consequences. He remained asymptomatic for more than 1 year of follow-up.

DISCUSSION

POTS/IOTS is characterised by increase in heart rate of more than 30/min associated with orthostatic symptoms of inadequate cerebral perfusion and increase in levels of catecholamine on standing in absence of orthostatic hypotension.^{1–4}

Literature reveals other descriptions for this condition such as soldier's heart, neurocirculatory asthenia and MV prolapse syndrome.⁵

Most patients with orthostatic intolerance are female aged 20–50 years. Symptoms of POTS are similar to chronic fatigue syndrome in many respects.⁵

Various symptoms described by POTS patients include: fatiguability, light-headedness, clouding of thoughts, blurred vision, anxiety, shoulder and neck pains, nausea, diaphoresis, dizziness, palpitations, chest discomfort, exercise intolerance and frank syncope.^{4 6}

In one of the study done for POTS subjects, fatigue was commonest symptom; other patients reported syncope with prolonged standing (39%) which the patients learned to prevent by sitting or lying down. Significant number of POTS patients were also found to have mitral valve prolapse (46%) and irritable bowel syndrome (23%).⁶

In normal subjects, plasma norepinephrine rises after standing in first 5 min, in POTS this continues to rise for 30 min. This is attributed to reduced clearance of norepinephrine. A possible mechanism suggested for reduced clearance of noradrenaline is decreased cardiac output secondary to hypovolemia or reduced venous return.⁴

Renin angiotensin axis

It is postulated that POTS patients have reduced plasma volume, inappropriately low levels of rennin and aldosterone (which promote sodium retention and increase plasma volume). These patients were also found to have low RBC mass. These findings put together suggests that renal abnormalities may be crucial in POTS pathophysiology.⁷

Apart from hypovolemia in POTS, there is possibility of partial sympathetic dysautonomia. It is not clear if dysautonomia is functional or anatomical. It is also not clear if primary abnormality is one of peripheral autonomic function or central regulatory mechanism.⁴

The diagnosis of POTS is made with head tilt table testing. A positive result is a sustained heart rate increase of greater than 30 bpm or an increase to 120 bpm or greater within the first 10 min of tilt. In addition to tilt table testing, other investigations include catecholamine levels. Various authors have suggested different levels of norepinephrine levels in standing. But most agree on levels 720.

Management of POTS

As patients with POTS may have undergone a long period of non-specific symptoms and a battery of investigations before coming to diagnosis, it is important to have proper counselling, explain the diagnosis and management plan to the patient.

It is equally important to explain the benign nature of the condition.

One of the first line of treatment suggested is increase in water intake in reducing symptoms and postural tachycardia.

Compressive stockings extending to the waist to reduce pooling of blood in the lower limbs and splanchnic circulation have shown benefits in reducing symptoms of POTS.

Increase in oral salt intake has been shown to have good effect on symptoms and tachycardia.

Drugs used for POTS are:

1. Fludrocortisone by retaining sodium and water and thereby expanding plasma volume helps reduce signs and symptoms.
2. Midodrine, an adrenoceptor agonist is useful due to its vasoconstrictor properties.
3. As POTS is a hyperadrenergic state, β blockers also have been used successfully.
4. Other treatment options for POTS include selective serotonin reuptake inhibitors which reduce venous pooling through stimulation of the standing vasoconstriction reflex. Other potential therapies include

pyridostigmine (an acetylcholine esterase inhibitor) and erythropoietin.

Learning points

- POTS is a disabling condition affecting young, but otherwise healthy women more than men.
- It is important to rule out other cardiac, endocrine causes leading to similar signs and symptoms.
- Other diagnoses such as autonomic neuropathy, vasovagal syncope or chronic fatigue syndrome should also be ruled out using standard diagnostic tools.
- The diagnosis of POTS is straightforward, that is, demonstrating tachycardia on tilt table testing and norepinephrine levels. However, due to lack of awareness of this relatively benign condition, it can be easily missed.

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Competing interests None.

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