# Odd turns in adult life: voltage-gated potassium channel antibody syndrome

## Introduction

Voltage-gated potassium channel complex antibody syndrome is an unusual cause of autoimmune encephalitis. Patients usually present in later life with adult onset seizures, behavioural and memory changes, and occasionally psychiatric symptoms.

## Discussion

Voltage-gated potassium channel antibody syndrome is an unusual cause of autoimmune encephalitis and/or peripheral nervous system effects. It is frequently associated with hyponatraemia, considered likely to be secondary to inappropriate antidiuretic hormone secretion (Vincent et al, 2004) and contributory to any cerebral dysfunction.

Patients usually present in later life with adult onset seizures, behavioural and memory changes, insomnia and occasionally psychiatric symptoms (Parthasarathi et al, 2006). CNS features range widely and can include apathy, depression, insomnia, memory loss, absence-type attacks, agitation and confusion. Peripheral nervous system symptoms include stiffness, fasciculations and hyperhydrosis. Previously considered a uniquely paraneoplastic phenomena, and typically associated with small cell lung cancer, thymoma, testicular and breast cancer, it is now appreciated that cases do occur in the absence of malignancy (Irani et al, 2010).

This patient exhibited a combination of central (absence attacks, dizziness and sleep disturbance) and autonomic (syncope, sweating and tachycardia) nervous system symptomatology but an absence of hyponatraemia or overt seizure activity. Hyponatraemia is not obligatory (Barajas et al, 2010), and while cases with autoimmune limbic encephalitis frequently demonstrate magnetic resonance imaging changes within the temporal lobes and hippocampal regions (Vincent et al, 2004; Merchut, 2010; Toyota et al, 2014), this is not universal.

Management of this syndrome involves immunosuppression with corticosteroids or steroid-sparing agents, and on occasions plasma exchange or intravenous immunoglobulin. These therapies also appear to address the associated hyponatraemia (Misawa and Mizusawa, 2010). A sub-type of the disorder, termed Moran syndrome, involves the peripheral nervous system (neuromyotonia), autonomic (hyperhidrosis, constipation, incontinence and cardiac arrhythmias) and central nervous systems (insomnia, hallucinations, memory impairment and seizures) (Misawa and Mizusawa, 2010).

## Conclusions

Voltage-gated potassium channel complex antibody syndrome, this infrequent autoimmune disorder, needs to be considered in adult patients presenting with subacute onset neurological dysfunction, atypical seizures or behavioural change, not least because it is amenable to therapy. BJHM

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### CASE REPORT

A 66-year-old Caucasian man reported having had 'odd turns' over the preceding 15 years. Having suffered a myocardial infarction 12 years ago it was felt that these events might be cardiac in origin. Exercise treadmill stress testing, Holter electrocardiography, an implanted Reveal device and ambulatory 24-hour blood pressure monitoring were all normal. Longstanding medications were losartan (50 mg daily), doxazosin (8 mg daily), bisoprolol (1.25 mg daily), aspirin (75 mg daily) and simvastatin (40 mg daily).

On most occasions these episodes would occur when he was inactive and frequently in the middle of the night. Each episode would last a few minutes and be associated with dizziness and variable feelings of discomfort in his face or sometimes limbs. He would always feel unwell and would have the sensation that he would faint. On several occasions he was syncopal and attended the accident and emergency department where no abnormality was found. He was described as appearing 'pale and sweaty' during an episode and was usually mildly tachycardic.

A magnetic resonance imaging scan of the brain revealed an incidental intracranial posterior fossa arachnoid cyst. The past history included headaches and retro-orbital discomfort, diagnosed as migraine and responding to the use of beta-blockers following his cardiac event. An electroencephalogram, with intermittent photic stimulation, showed no evidence of a focal seizure disorder. CSF examination revealed a mildly elevated immunoglobulin G, that was not oligoclonal. Blood chemistry, including sodium, calcium, folate and vitamin $B_{12}$, was normal.

The frequency of symptoms increased to almost daily episodes lasting around 30 seconds with a spreading sensation of warmth, occasional dizziness, visual disturbance and a sensation of impending syncope. He was commenced on lamotrigine at 50 mg twice daily with a partial response to this 'migraine-like' syndrome.

The results of a voltage-gated potassium channel antibody titre revealed an elevated level (151 pmol/litre, normal range 1–100 pmol/litre). Immunosuppressive therapy was commenced with mycophenolate mofetil 500 mg twice daily, with a significant improvement in symptomatology, with the intention of escalating the dose to abolish his symptoms. A subsequent whole body positron emission tomographic scan, performed to rule out malignancy, was normal.

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## LEARNING POINTS

- Anti-voltage-gated potassium channel antibody-associated disorders should be considered in unexplained adult onset neurological disorders.
- Anti-voltage-gated potassium channel antibody-associated disorders respond to treatment with immunosuppressive drugs.
- Malignancy should be excluded as voltage-gated potassium channel antibody complex disorder may represent a paraneoplastic disorder.



# Congenital absence of inferior vena cava: an overlooked cause of deep vein thrombosis

A 28-year-old woman presented with pain and numbness around the sacral area which was attributed to musculoskeletal problems. She re-presented with increasing right leg swelling in the next 2 days, confirmed to be an extensive femoral thrombus on ultrasound Doppler. Computed tomography venogram, in preparation for thrombolysis, demonstrated absent iliac veins and lower inferior vena cava (*Figure 1*). Thrombolysis and anticoagulation resulted in significant symptomatic improvement. She was discharged on lifelong anticoagulation.

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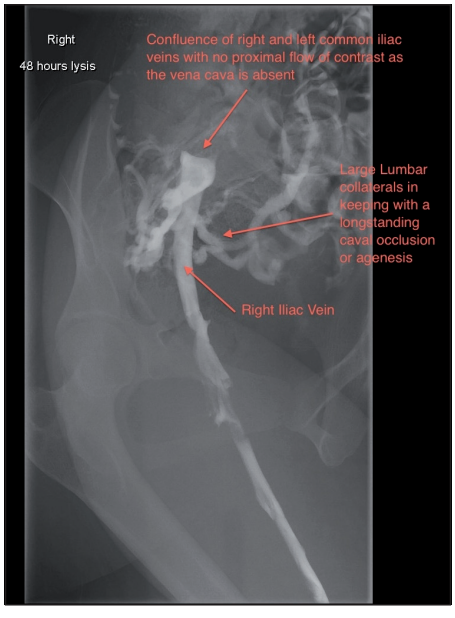
Congenital absence of inferior vena cava is present in 0.3–0.5% of the population with a higher prevalence in individuals with cardiovascular problems (2%) (Bami et al, 2015). About 5% of patients under 30 years of age presenting with unprovoked deep vein thrombosis are found to have absent inferior vena cava, possibly as a result of embryological dysgenesis and perinatal thrombosis (Lambert et al, 2010; Parsa et al, 2015). Collateral flow which develops is insufficient, risking deep vein thrombosis development, although smaller vessels means reduced risk of pulmonary embolism. Diagnosis requires computed tomography or magnetic resonance angiography (Dougherty et al, 1996). Treatment is with anticoagulation and thrombolysis with low bleeding risk. **BJHM**

**Figure 1. Computed tomography venogram demonstrating the absence of the inferior vena cava.**

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