Learning Point for Clinicians
Voltage gated potassium channel (VGKC) encephalitis is a recently recognized autoimmune condition with antibodies against components of the VGKC protein complex. Clinical phenotype can vary, but patients typically present with a limbic encephalitis involving amnesia, seizures, psychiatric disturbance and occasionally faciobrachial dystonic seizures.

Case report

A 66-year-old man presented following a generalized tonic-clonic seizure and a 1 month history of paroxysmal episodes with autonomic and sensory symptoms. Magnetic resonance imaging (MRI) found right medial temporal lobe swelling and high signal. Cardiac telemetry found supraventricular tachycardia and prolonged pauses during the paroxysmal episodes. Voltage gated potassium channel (VGKC) antibodies were positive. Immunomodulatory treatment lead to resolution of symptoms, MRI and cardiac abnormalities. This case highlights a recently recognized and treatable cause of late onset epilepsy and cardiac arrhythmia—typical presentations to primary care or the acute medical take. In our patient, an incorrect initial diagnosis of brain tumour and unnecessary permanent pacemaker (PPM) were avoided after a unifying diagnosis of VGKC limbic encephalopathy was made and treated.
Cardiac electrophysiological studies suggested significant bundle of His disease; insertion of a PPM was considered.

An alternate diagnosis of VGKC limbic encephalitis—explaining cerebral and cardiac manifestations—was confirmed with VGKC-complex antibodies positive at 1528 pM (0–100 pM), LGI1 subtype. No underlying malignancy was found on whole body CT or positron emission tomography (PET) scanning.

Treatment with intravenous immunoglobulins (IVIg) and methylprednisolone (1 g IV for 3 days and tapering dose of prednisolone), led to resolution of all symptoms. A 24-h electrocardiogram (ECG) 4 months later demonstrated sinus rhythm with occasional ventricular ectopics and bigeminy but no arrhythmias. There has been no recurrence at 1 year and repeat VGKC-complex antibody was negative. Follow-up MRI brain scan demonstrates resolution of the medial right temporal lobe high signal.

Discussion

Our patient was first diagnosed with a brain tumour and then cardiac dysrhythmia and was heading for pacemaker insertion before his VGKC-complex antibody encephalitis was identified and successfully treated; with parallel resolution of his cardiac problems. This case, as far as we are aware, is the first that describes symptomatic cardiac arrhythmia in association with limbic encephalitis with VGKC-complex antibodies. These antibodies bind to components of a protein complex associated with VGKCs. Leucine-rich, glioma-inactivated 1 (LGI1) is the most common target and VGKC-complex/LGI1 antibodies can also be found in patients with frequent short-lived faciobrachial dystonic seizures. MRI changes in limbic encephalitis typically show mesial temporal and amygdala high signal and swelling that can be mistaken for low grade glioma or cortical dysplasia but resolve over time, or result in mesial temporal atrophy. The paroxysmal episodes in our patient were frequent and short lived and associated with cardiac arrhythmia on 24-h ECG and resolved with immune-modulatory treatment. Focal epileptic seizures, in particular, from non-dominant mesial temporal structures are associated with cardiac arrhythmia; in our patient bundle of His disease on electrophysiological studies and echocardiogram changes being attributable to longstanding hypertension. Alternatively, symptomatic cardiac arrhythmias may have been due to a direct effect of VGKC-complex/LGI1 antibodies on the myocardium. Autoimmune limbic encephalitis is a new and exciting area that has implications in understanding hitherto unknown pathophysiology of a number of neurological presentations.

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Conflict of interest: A.V. and the University of Oxford hold patents and receive royalties for antibody assays.

References


