Supplementary Table

Supplementary Table S1 Investigation results

Initial Investigations	Findings	
Hemoglobin	11.2 g/dL	
WBC	13.15 $\times 10^9$ /L	(Peak: 32 $\times 10^9$ /L on Day 10)
Platelet	100 $\times 10^9$/L	(Trough: 60 $\times 10^9$/L on Day 2)
INR	1.25	(Peak: 1.7 on Day 6)
Creatinine	181 mmol/L	(Peak: 241 mmol/L on Day 2)
ALT	80 U/L	(Peak: 678 U/L on Day 6)
hsTnI	88,874 ng/L	(Peak: 88,874 ng/L on Day 1)
CK	108,510 U/L	(Peak: 614,400 U/L on Day 5)
LDH	770 U/L	(Peak: 14,363 U/L on Day 6)
CRP	45 mg/L	(Peak: 111 mg/L on Day 4)
ESR	23 mm/h	(Peak: 47 mm/h on Day 4)
Ferritin	10,536 pmol/L	
D-dimer	>10,000 ng/mL	
Blood gas	pH: 7.39, pCO_2: 3.3 kPa, HCO_3: 14.7 mmol/L, BE: −9.1 mmol/L	
Lactate	3.1 mmol/L	
Procalcitonin	31.57 ng/mL	
Urine myoglobin	Positive	
CT brain	Normal	
Echocardiogram	Mildly impaired contractility with some dyssynchrony of wall motion	
Cerebrospinal fluid		
WBC	10/mm3	
Protein	0.18 g/L	
Culture, viral study	Negative	
Oligoclonal band, IgG	Negative	
Anti-NMDA-receptor, anti-CASPR2, anti-LGI1, anti-AMPAR1/2, anti-DPPX, anti-GABAR B1/B2 antibodies	Negative	
Autoimmune workup		
Anti-GAD antibody (cutoff: 10 IU/mL)	170 IU/mL (positive)	
ANA	160 (high)	
Anti-Ro antibody	Positive	
Anti-dsDNA, anti-La, anti-Sm	Negative	
Anti-MOG, anti-aquaporin 4 antibody, anti-NMDA-receptor antibody	Negative	
Anti-thyroglobulin, anti-thyroid peroxidase antibody, anti-gastric parietal cell antibody	Negative	
Microbiological workup		
Cultures from blood, urine, endotracheal tube, throat swab, stool	Negative	
Serum Japanese encephalitis virus, dengue virus, EBV serology	Negative	
Metabolic workup		
Dried blood spot test	Normal	
Urine metabolic profile	Normal	
Acylcarnitine profile	Generalized elevation of short chain acylcarnitines	

(*Continued*)

Initial Investigations	Findings
Toxicological workup	
Urine toxicology	Negative
Tumor markers	
AFP, CEA	Normal
Others	
Fasting glucose, HbA1C	Normal
Vitamin B12	Normal
Thyroid function	Normal
Imaging	
MRI brain	Subtle hyperintensity in bilateral medial temporal lobes on T2 and FLAIR sequence (Fig. 1)
MRI spine	Mildly swollen spinal cord with bilateral T2 hyperintense signal extending from T10/11 to conus medullaris (Fig. 1)
PET-CT	Symmetrical diffuse prominent activity in thyroid
VCUG	Neurogenic bladder
USG abdomen	Normal
Colonoscopy	Normal
Electrophysiological studies	
EEG	Generalized delta brush waves
EMG	Difficult to assess due to patient's uncooperativeness

Abbreviations: AFP, α-fetoprotein; ALT, alanine transaminase; ANA, antinuclear antibody; anti-AMPAR1/2, anti-α-amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid receptor; anti-DPPX, anti-dipeptidyl-peptidase-like protein 6; anti-CASPR2, anti-contactin-associated protein-like 2; anti-dsDNA, anti-double-stranded DNA; anti-GABAR B1/B2, anti-gamma-aminobutyric acid B1/B2; anti-GAD, anti-glutamic acid decarboxylase; anti-LGI1, anti-leucine-rich glioma inactivated 1; anti-MOG, anti-myelin oligodendrocyte glycoprotein; anti-NMDA-receptor, anti-N-methyl-d-aspartate-receptor; anti-Sm, anti-smith; BE, base excess; CEA, carcinoembryonic antigen; CK, creatine kinase; CRP, C-reactive protein; CT, computed-tomography; EBV, Ebstein Barr virus; EEG, electroencephalogram; EMG, electromyogram; ESR, erythrocyte sedimentation rate; HbA1C, hemoglobin A1C; hsTnI, high-sensitivity troponin I; IgG immunoglobulin G; INR, international normalized ratio; LDH, lactate dehydrogenase; MRI, magnetic resonance imaging; PET-CT, positron emission tomography–computed tomography; USG, ultrasound; VCUG, voiding cystourethrogram; WBC, white blood cell.

Supplementary Table S2 Diagnostic criteria for progressive encephalomyelitis with rigidity and myoclonus 1

Major criteria	• Clinical presentation including typical body regions involved (neck, torso, extremities, brainstem, and/or cerebellar symptoms) • Hallmark triggers for spasms/increased rigidity (abrupt loud noises, cold weather, open spaces, emotional stress, and/or tactile stimuli) • Exam findings: admixture of other phenotypes findings plus encephalopathy and severe torso rigidity and/or myoclonus (multifocal or generalized) • Presence of serum autoantibody to GAD65 (high titer), glycine receptor, or amphiphysin • EEG (generalized slowing and/or epileptic discharges) • Exclusion of alternative diagnoses and no better explanation for syndrome
Minor criteria	• Autonomic dysfunction • Presence of CSF autoantibody to GAD65, glycine receptor, or amphiphysin • CSF pleocytosis • CSF-restricted oligoclonal band • EMG demonstrating co-contraction of agonist and antagonist muscles and/or continuous motor unit activity in affected muscles (paraspinal/abdominal musculature and/or legs/arms) • Brain MRI demonstrating T2/contrast-enhancing lesion(s) in brainstem • Brain FDG-PET demonstrates hyper- or hypometabolism within cortices
Interpretation	• Meets all major and minor criteria = Definitive diagnosis • Meets all major criteria and no minor criteria = Definitive diagnosis • Meets four major criteria (must include serum autoantibody and exclusion of alternative diagnoses and no better explanation for syndrome) and at least two minor criteria = Definitive diagnosis • Meets three major criteria (must include serum autoantibody and exclusion of alternative diagnoses and no better explanation for syndrome) and at least two minor criteria = Definitive diagnosis • Meets three major criteria (must include exclusion of alternative diagnoses and no better explanation for syndrome) and at least two minor criteria = Probable diagnosis • Meets three major criteria (must include exclusion of alternative diagnoses and no better explanation for syndrome) and less than two minor criteria = Possible diagnosis • Meets two major criteria (must include exclusion of alternative diagnoses and no better explanation for syndrome) and at least two minor criteria = Possible diagnosis

Abbreviations: CSF, cerebrospinal fluid; EEG, electroencephalography; EMG, electromyography; FDG-PET, fluorodeoxyglucose-positron emission tomography; GAD65, glutamic acid decarboxylase 65 kDa isoform; MRI, magnetic resonance imaging.

