Table e1. Serological findings in 221 definite AE cases

Category	Ab	N
Limbic encephalitis, 127	*Lgi1[a]	53
	Negative[b]	24
	GAD65	18
	*CASPR2	**7**
	ANNA-1 (anti-Hu)	**5**
	Ma2	6
	AGNA (anti-SOX-1)[c]	5
	*AMPA-R	3
	*$GABA_BR$[d]	**2**
	CRMP-5	1
	*DPPX	1
	AK5	1
	*$GABA_AR$[e]	1
NMDA-R, 32	*NMDA-R[f]	32
ADEM, 8	*MOG	4
	Negative	4
Other, 54	*Lgi1	23
	GFAP	9
	*DPPX	6
	GAD65	3
	ANNA-1 (anti-Hu)	2
	CRMP-5	2
	AGNA (anti-SOX1)	1
	*AMPA-R	1
	Amphiphysin	1
	*CASPR2	1
	*MOG	1
	Ma2	1
	*mGluR5	1
	*$GABA_AR$[g]	1
	NIF[h]	1

* = antibody directed against neural cell surface antigen (non-asterisked include those antibodies against intracellular antigens; a, 1 had coexisting CASPR2-IgG; b, 4 had unclassified Abs detected; c, 1 had coexisting $GABA_BR$ Ab; d, 1 had coexisting ANNA-1 and amphiphysin antibody; e, with coexisting high-titer GAD65 antibody; f, 2 with coexisting GFAP-IgG, 1 with coexisting MOG-IgG; g, 1 had coexisting high-titer GAD65 antibody; neuronal intermediate heavy chain and light chain (NF-H, NF-L) Abs were the NIF Ab subtypes detected. AGNA = anti glial/neuronal nuclear antibody; AK5 = adenylate kinase 5; AMPA = α-amino-3-hydroxy-5-methyl-4-isoxazole propionic acid; Amphi = amphiphysin; ANNA = antineuronal nuclear antibody; CASPR= contactin-associated protein; CRMP = collapsin-response mediator protein; DPPX = dipeptidyl peptidase; GABA = gamma amino butyric acid; GAD65 = glutamic acid decarboxylase 65

kDa isoform; GFAP = glial fibrillary acidic protein; Lgi1 = leucine rich glioma inactivated 1; NIF = neuronal intermediate filament antibodies; MOG = myelin oligodendrocyte glycoprotein; NMDA = n-methyl-D-aspartate; R = receptor; SOX1 = SRY-box transcription factor 1.

Table e2 Diagnostic and therapeutic features of 42 patients not meeting at least possible AE criteria, diagnosed with AE-IgG seronegative autoimmune encephalopathies at Mayo Clinic.

Study No/Age/Sex/ Subacute onset	Memory loss, altered mental status or psych	New focal CNS findings	Seizures	Supportive MRI Brain	CSF (*)	Autoimmune disease	Other notable features	Non-AE AutoAb clues	Immune-therapy response	Objective changes reported
196/34/F/Y	Y	N	N	N	N	Hypothyroid, B12 deficiency	-	N	Steroids	Resolution of AMS & diffuse delta slowing on EEG
197/63/M/Y	Y	N	N	N	N	Hypothyroid, celiac	-	N	Steroids, PLEX	Resolution of aphasia, AMS & fluctuating hemiparesis, & theta slowing on EEG.
199/13/F/Y	Y	N	N	N	N	Hypothyroid	-	N	IVIg, rituximab	Improved matrix reasoning & math calculation on neuropsycho-metric testing
201/32/F/Y	Y	N	N	N	N	Graves disease, sclerosing cholangitis, UC	-	N	Steroids	Encephalopathy resolved
205/83/M/Y	Y	N	N	N	N	Hypothyroid, B12 deficiency	-	N	Steroids	From coma to Kokmen of 34/38
211/54/F/Y	Y	N	N	N	N	-	-	N	Steroids	Resolution of encephalopathy & delta slowing on EEG
303/38/M/Y	N	N	N	Y	N	-	Brainstem symptoms, headache, ADEM-like MRI	N	Steroids	Resolution of clinical syndrome & MRI findings

313/54/M/Y	N	N	N	Y	PI	-	Episodic weakness R hand & foot, dysarthria. Normal EEG	N	Steroids	Resolution of dysarthria
316/20/M/N	Y	N	N	N	P	-	-	N	Steroids	PET-CT global hypometabolism resolved
322/25/F/Y	N	N	Y	Y	N*	-	Right sided weakness & aphasia, L sided encephalitis on MRI	N	Steroids	Resolution of clinical syndrome & MRI findings
323/61/M/Y	Y	N	N	N	N	-	CSF protein 282mg/dL, myeloma	N	Steroids	Improvements in manual speed & dexterity, spatial ability, cognitive speed & efficiency, working memory, & verbal fluency.
330/11/F/Y	N	Y	N	N	N	-	Post infectious encephalo-myelopathy (cognitive, dystonia, myelopathy)	N	PLEX, rituximab	Improved cognition
334/43/M/N	Y	N	N	Y	P	-	Gastric & neuroendocrine cancer history	N	Steroids	Improvement across the neuropsychometric testing battery

335/49/F/N	Y	N	N	N	O	RA	Intermitted word finding difficulty & ataxia	N	IVIg, mycopheno late	Kokmen score 33/38 to 38/38.
336/41/M/Y	Y	N	N	N	N	Graves' disease	CSF protein 105	N	Steroids	Improved neuropsychometric scores
339/73/F/Y	Y	N	N	N	O	-	Bladder & ovarian carcinomas by history, checkpoint inhibitor therapy	N	Steroids, IVIg	MOCA improved 18/30 to 28/30, worsened with taper
340/58/M/Y	Y	N	N	N	N	Autoimmune hemolytic anemia, ITP	-	GAD65LT	Steroids	Encephalopathy improved, worsened with taper
343/45/F/Y	Y	N	N	N	N	Type 1 DM, lupus nephritis	-	GAD65LT	IVIg	Improved performances on tasks of psychomotor speed, verbal fluency, constructional praxis, acquisition & retention of verbal information.
344/39/F/Y	Y	N	N	N	N	Hashimoto thyroiditis	Ovarian teratoma	GAD65LT	Steroids	Encephalopathy improved, worsened with taper

356/72/M/Y	Y	N	N	N	N	-	Metastatic renal cell carcinoma, encephalopathy with disinhibition, personality changes	GAD65LT	Steroids, PLEX	Encephalopathy resolved
357/71/F/N	Y	Y	N	Y	N	Hypothyroid	-	N	Steroids	Improved encephalopathy, residual amnesia, EEG & MRI changes
360/54/F/N	Y	Y	N	N	O	-	--	p-ANCA	Steroids, cyclophosphamide	Improved cognition with steroids.
364/50/M/N	Y	Y	N	N	N	-	-	Phospholipid	Steroids, rituximab	Improved language function
375/51/F/Y	Y	N	N	N	N	Hypothyroid	-	Thyroglobulin	Steroids	Improved cognitive scores
378/60/M/Y	N	Y	N	N	PO*	-	Progressive unsteadiness, tremor, nonspecific cognitive symptoms, dysgeusia	TPO	Steroids, myco-phenolate	Cognitive scores, MRI, EEG normalized
383/53/M/Y	Y	N	N	N	N	-	Extensive family history of autoimmune disease (thyroid, type 1DM) in multiple 1st degree family members	TPO	Steroids	Significant but incomplete, improvement on neuropsycho-metric testing

385/67/M/N	Y	N	N	N	P	-	-	TPO	Steroids, IVIg, PLEX	Resolution of paranoia & hallucinations (3 episodes)
386/51/F/Y	Y	N	N	N	N	LES	-	TPO	Steroids	Improved neuropsychiatric disorder
390/46/F/Y	N	Y	N	Y	POI	Celiac disease	Ataxia, dysarthria, nonspecific cognitive symptoms, & upper motor neuron signs	TPO	Steroids	Kokmen STMS score improved from 29/38 to 38/38, improved ataxia
392/16/F/N	Y	N	Y	N	P	Hypothyroid	-	TPO	Steroids	Encephalopathy & R temporoparietal epileptiform EEG discharges resolved.
394/42/F/Y	Y	N	N	N	N	Hypothyroid	-	TPO	Steroids, IVIg	Improved encephalopathy with IVIg
396/58/M/N	Y	Y	N	N	I	-		TPO	Steroids	Kokmen STMS score improved from 16/38 to 26/38
397/34/F/Y	N	Y	N	Y	PO	-	Dizziness, double vision, slurred speech, and gait ataxia	TPO	Steroids	Improved ambulation (wheelchair to cane)
398/49/F/Y	Y	N	N	N	N	Hypothyroid	-	TPO	Steroids	Resolution of stroke-like episodes

399/29/F/N	N	Y	N	N	N	Hypothyroid, B12 deficiency, Type 1 DM	Light sensitivity R arm tremors, facial twitching, stroke like episode	TPO	Steroids	Resolution of encephalopathy and L temporal slowing on EEG
400/48/M/Y	Y	N	N	N	N	Hypothyroid	-	TPO	Steroids	Encephalopathy resolved
401/40/F/Y	Y	N	N	N	N	Leukocytoclastic vasculitis	-	TPO, thyroglobulin	Steroids	Encephalopathy resolved
402/72/F/Y	Y	N	N	N	N	B12 deficiency	-	TPO, thyroglobulin	Steroids	Encephalopathy resolved
463/21/F/N	Y	N	N	Y	N	-	Fever, cough, stiff neck, MRI changes of ADEM.	TPO	Steroids	Clinical meningitis resolved, some intermittent headaches
478/57/M/N	Y	Y	N	N	P	-	-	Unclassified neural	No	No change
537/34/F/N	Y	N	N	Y	N	-	Premature ovarian failure, type 1 diabetes thyroid disease in family	N	IVIg	Improved neuropsychometric scores
538/22/F/N	Y	N	N	N	N	-	-	GAD65LT, TPO	Steroids	Improved encephalopathy

ADEM=acute disseminated encephalomyelitis; AE=autoimmune encephalitis; AMS = altered mental status; AutoAb = autoantibody; DM = diabetes mellitus; EEG = electroencephalogram; F=female; GAD65LT = low titer (< 20 nmol/L or < 10,000 IU/mL) glutamic acid decarboxylase, 65 kilodalton isoform antibody; ITP=immune thrombocytopenia; IVIg = intravenous immune globulin; L=left; M=male; MOCA=Montreal cognitive assessment; p-ANCA= anti neutrophil cytoplasmic antibody, p-type; PLEX = plasma exchange; R=right; Y=yes; N=No; STMS = short test of mental status; TPO = thyroperoxidase; UC=ulcerative colitis.