Clinical and MRI features in Southern Italy cases of Autoimmune Epilepsy

S. G. Quitadamo, M. Tappatà, C. Tortorella, A. Introna, N. Pilolli, T. Francavilla, A. La Neve, I. Simone

Department of Basic Medical Sciences, Neurosciences and Sense Organs - University of Bari

Objectives: Several studies reported a high prevalence of serum/CSF neural autoantibodies (Abs) in epileptic patients (1,2). The APE Score, including clinical/MRI/CSF features, has been recently proposed to predict the probability of autoimmune etiology in epilepsy. An APE score cut-off of 4 has been selected to indicate patients to Abs testing (3). The aims of this study is to retrospectively evaluate the APE Score in our patients and to correlate the positivity for neural Abs to clinical/MRI features.

Materials and Methods: Six epileptic patients (5 females, 1 male; age range 16-57 years) were evaluated. Abs panels were tested by commercial kits.

Results: We examined 6 patients referring to the Centre of Epilepsy of our Department for onset of epilepsy of undefined etiology. Routine CSF was normal and MRI findings excluded infective or tumoral etiologies. Four patients showed Abs (3 anti-GluR3, 1 anti-LGI1) in CSF and 2 (1anti-Hu, 1 anti-NMDAR) in serum. The range of APE Score was 1-4 but only in one patient it reached the suggested cut-off of 4 (Table 2).
Drug-resistance was present in 2 patients and 2 had seizure-relapse trying to interrupt therapy. In 5/6 patients MRI was abnormal, in 3 suggestive of encephalitis with involvement of various area and one patient had mesial temporal sclerosis. Two patients had seizures in clusters and other two status epilepticus. Three patients underwent immunotherapy (ivIG/Steroid), one of them started long-term azathioprine because of severe disease course (Table 3).

Table 1. Components of the APE Score

Clinical Feature	Value[a]
New-onset, rapidly progressive mental status changes of 1-6 weeks, or new-onset seizure activity	1
Neuropsychiatric changes; agitation, aggressiveness, emotional lability	1
Autonomic dysfunction (presenting as labile blood pressure, labile heart rate, persistent tachycardia, postural hypotension)	1
Viral prodrome (runny nose, sore throat, low-grade fever), only to be scored in the absence of underlying malignancy	2
Facial dyskinesias or faciobrachial dystonic movements	2
Seizure refractory to at least 2 antiseizure medications	2
CSF findings consistent with inflammation (elevated CSF protein level >50 mg/dL and/or lymphocytic pleocytosis >5 cells/dL, if the total number of CSF RBCs is <1000 cells/dL)[b]	2
Brain MRI showing signal changes consistent with limbic encephalitis (medial temporal T2/FLAIR signal changes)[b]	2
Presence of underlying malignancy (excluding cutaneous squamous cell or basal cell carcinomas)	2
Total	15

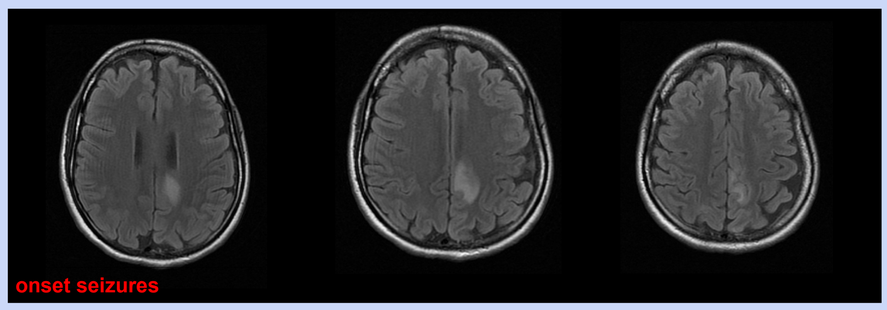
Table 2. Total APE score in our patients

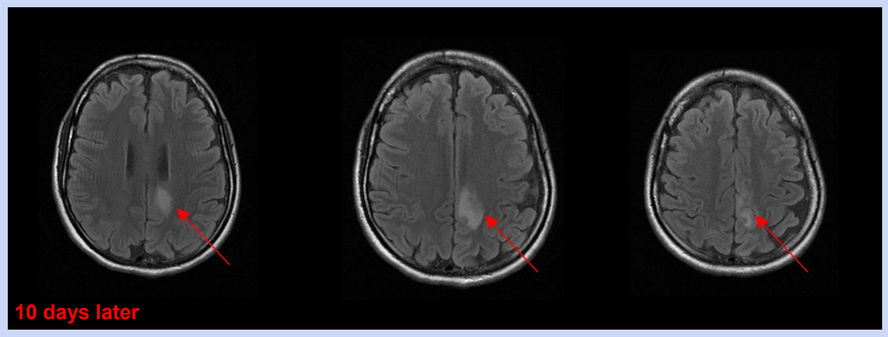
	GLUR3 (♀ 44y)	LGI1 (♀ 55y)	Hu (♀ 16y)	GLUR3 (♀ 57y)	NMDAR (♀ 19y)	GLUR3 (♂ 48y)
New-onset mental status/seizures	1	1	-	1	1	-
Neuropsychiatric	-	-	1	-	1	1
Autonomic	-	-	-	-	-	-
Viral prodrom	-	-	-	-	-	-
Facio-brachial	-	-	-	-	-	-
Seizure refractory to 2 medications	-	-	2	2	2	-
Inflammatory CSF	-	-	-	-	-	-
Limbic encephalitis	-	-	-	-	-	-
Malingnancy	-	-	-	-	-	-
Total APE score	1	1	3	3	4	1

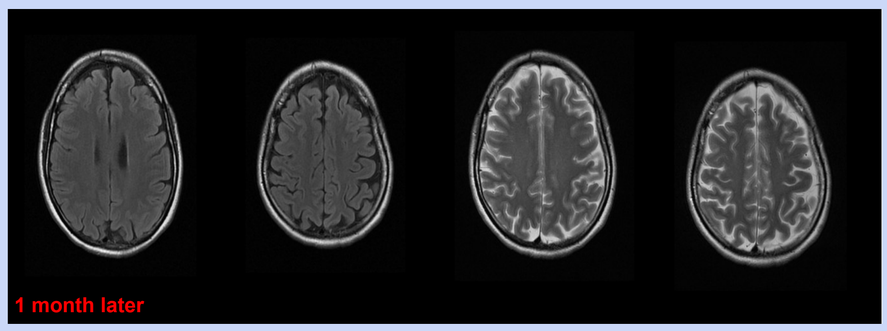
Table 3. Clinical and MRI features

	GLUR3 (♀ 44y)	LGI1 (♀ 55y)	Hu (♀ 16y)	GLUR3 (♀ 57y)	NMDAR (♀ 19y)	GLUR3 (♂ 48y)
Resistance (R) or Recurrence (RR)	-	RR	R	R	R	RR
MRI findings	Frontal	Normal	Cerebellar atrophy	Multifocal	Normal	Temporal Sclerosis
Status Epilepticus (SE) or Clusters (C)	C	-	-	SE	SE	C
Therapy	LEV, **MP**	LEV	LEV, LTG, LCS, CBZ	LEV, VPA, **IVIG, MP**	MDZ,CBZ **IVIG, AZA,** PB, **Plasm.**	VPA

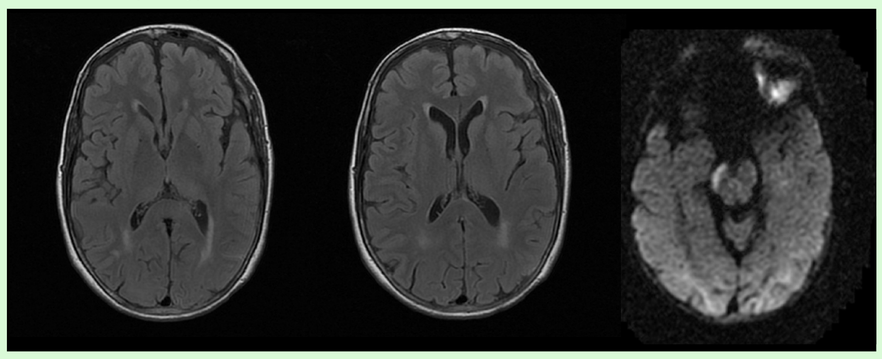
GLUR3 (♀ 44y)







GLUR3 (♀ 57y)



Multiple and asynchronous lesions (FLAIR/T2 and DWI)

Discussion and Conclusions: in our limited cohort, the APE-score was unable to identify the 83% of autoimmune epilepsy, then other clinical or MRI features should be considered to predict an autoimmune pathogenesis. CSF is important for differential diagnosis, but it may be often normal except for Abs presence. In relation to therapeutical intervention, testing Abs should be considered mainly for patients with drug resistance epilepsy of unknown etiology.

Quek AM et al. Autoimmune Epilepsy: clinical caracteristics and response to immunotherapy. Arch Neurol. 2012 May;69(5):582-93. 2012
Dubey D et al. Gaspard N. Autoimmune Epilepsy. Continuum (Minneap Minn). 2016 Feb;22(1 Epilepsy):227-45
Neurological Autoantibody Prevalence in Epilepsy of Unknown Etiology. JAMA Neurol. 2017 Apr 1;74(4):397-402

Sin
XLVIII CONGRESSO NAZIONALE
14-17 OTTOBRE 2017 – NAPOLI
WebPoster
http://congress.wooky.it/NEURO2017/