Seronegative Autoimmune Encephalitis with Evanescent Focal T2FLAIR Lesions

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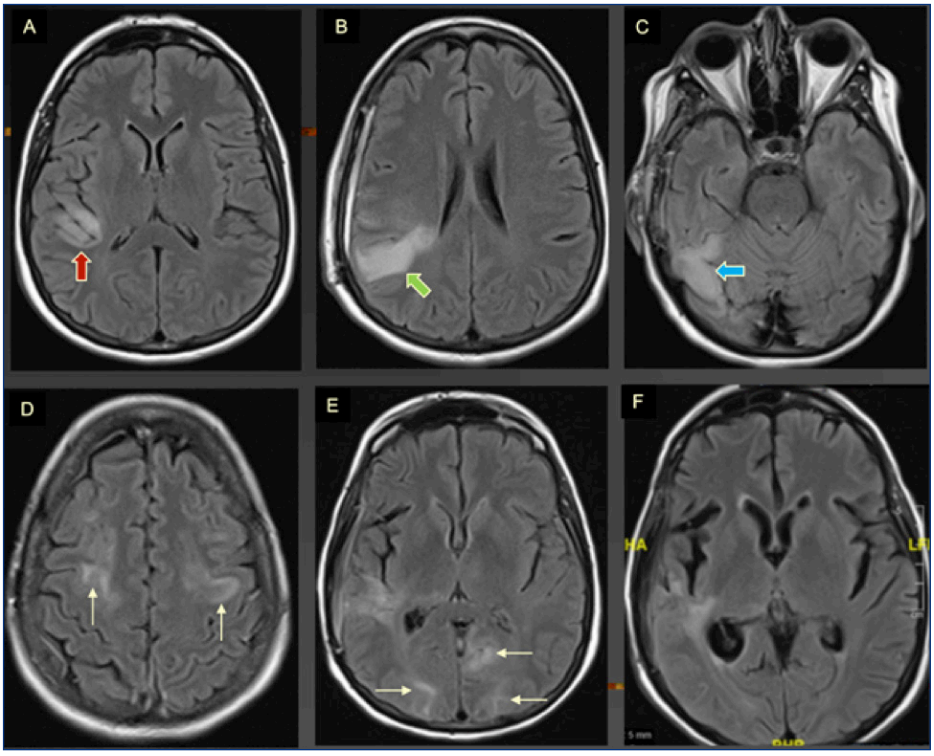
Autoimmune encephalitis is a misdirected immune response against self-antigens expressed in the central nervous system. The prevalence is estimated to be 13.7/100,000.[1] About half of the cases are seronegative. They do not have a recognized neural-specific IgG[2]. Typical autoimmune encephalitis causes rapid progression of new psychiatric symptoms (psychosis, aggression, catatonia, short-term memory deficits, or decreased level of consciousness) with one of the following: new focal CNS deficits, new seizures, CSF pleocytosis, or MRI features of encephalitis.[3]

Here, we present the MRI findings of evanescent focal T2FLAIR lesions in a 53-year-old female patient with seronegative autoimmune encephalitis who developed refractory epilepsy. Her past medical history was notable for plaque psoriasis, well controlled for six years with ustekinumab, an anti-IL-12 and anti-IL-23 monoclonal antibody which inhibits T-cell activation. She developed paroxysms lasting one to five minutes of seeing a spinning fan in her left upper visual field, of hearing indistinct voices, and of having left facial twitching, each sometimes associated with loss of consciousness. These were not recognized as seizures until five months after first seeing a spinning fan, when she crashed a vehicle due to a secondarily generalized seizure.

EEG at that time captured intermittent slowing over temporal regions (right greater than left) and occasional sharp waves in the right mid-temporal region. MRI showed a 2.5cm, non-enhancing, T1-hypointense and T2-hyperintense lesion in the right temporal lobe initially thought to represent a low-grade glioma (See **Figure 1A–F**). This was biopsied, and pathology demonstrated focal encephalitis without glioma. CSF showed pleocytosis with lymphocytic predominance (14 nucleated-cells), increased protein concentration of 73 mg/dl, and increased CSF-specific oligoclonal bands (12). Serum aquaporin-4 and myelin oligodendrocyte glycoprotein (MOG) antibody tests were negative. An infectious work-up including tests for HIV, JC virus, and a broad meningitis/encephalitis panel were negative. An autoimmune encephalitis cell-based immunofluorescence panel was negative.

Figure 1. Migratory and evanescent T2-FLAIR lesions associated with seronegative autoimmune encephalitis. [**A**] Five months after first seizure, focal lesion (red arrow) in right posterosuperior temporal lobe involving Heschl gyrus. Biopsy revealed "encephalitis." [**B**] One month later, a new lesion (green arrow) in cortex and white matter of the right inferior parietal lobule. Lesion in the right posterosuperior temporal gyrus had decreased in size (not shown). [**C**] Seven months after first seizure, another new lesion (blue arrow) in cortex and white matter of right posterior temporal lobe. [**D**,**E**] Nine months after first seizure, new subcortical white matter lesions (small arrows) in both posterior frontal and occipital lobes, resembling posterior reversible encephalopathy syndrome. [**F**] Ten months after first seizure, residual gliosis at site of initial lesion (compare to A). All other lesions had resolved but note interval brain atrophy with ventriculomegaly and wide Sylvian fissures.



While being evaluated, she had increasingly refractory seizures. Nine months after her first seizure she developed suicidal and homicidal thoughts, was found searching for a dog which did not exist, and she forgot the existence of her granddaughter. Shortly thereafter, she had a prolonged admission for status epilepticus. She was treated empirically with methylprednisolone 1250 mg daily for three days, followed immediately by IVIG 0.4 mg/kg over five days, followed two weeks later by methotrexate 1g monthly. At the time of receiving her first dose of rituximab she was minimally responsive and required a tracheostomy, a PEG tube, and five anti-epileptic drugs. Three months after her first dose she followed up as an outpatient and was living with her family, her tracheostomy was decannulated, she could eat by mouth, she could follow a conversation, and she could start to wean her antiepileptic drugs.

References

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Disclosures

The authors have no conflicts of interest relevant to this article to disclose.

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