SURVEY QUESTIONS: When do you suspect autoimmune encephalitis and what is the role of antibody testing?

Aravind Ganesh, MD, Luca Bartolini, MD and Sarah Wesley, MD MPH

CASE 1. A 20-year-old woman without past psychiatric history presents with delusions, agitation, and auditory hallucinations developing over the past 10 days. On examination, she often rambles incoherently and cannot name objects presented to her; otherwise neurological examination is normal. Lumbar puncture reveals slight lymphocytic pleocytosis but is otherwise unremarkable. Brain MRI shows T2 hyperintensity in the medial temporal lobes. Extended toxicology screening, CT chest/abdomen/pelvis and extensive infectious workup are all negative.

1. Would you send an autoimmune antibody panel at this point?

- a. Yes
- b. No

If answering yes to question #1 (skip logic):

1b. What type of panel would you send?

- Serum
- Serum and CSF

If answering yes to question #1 (skip logic):

1c. Autoantibody panel is pending and will take two weeks to return. Would you start treatment for presumed autoimmune encephalitis?

- Yes
- No, I would wait for the antibody panel to come back and would start only if positive at least in serum
- No, I would wait for the antibody panel to come back and would start only if positive in both serum and CSF
- Free text

If answering yes to question #1c (skip logic):

1d. After two weeks from initial treatment with first-line immunotherapy, the patient still does not make significant clinical improvement. Autoantibody panel comes back negative in both serum and CSF. What is your strategy at this point?

- Continue with immunotherapy (1st and/or 2nd line agents)
- Stop immunotherapy and reassess for other causes
- Free text

CASE 2. A 40-year-old man with type 1 diabetes mellitus presents to your hospital with new-onset seizures. Serum glucose is within range. On neurological exam, you observe no deficits other than mild short-term memory impairment. Over the course of the next week, he continues to have intermittent EEG-confirmed seizures arising from the right temporal lobe. High-resolution MRI brain shows T2 signal hyperintensity in the right hippocampus and no other abnormality. He has no history of febrile seizures and no family history of epilepsy. Extensive infectious workup is negative.

2. Would you send an autoantibody panel at this point?

- No
- Yes, in both serum and CSF
- Yes, in serum only
- Yes, in CSF only
- Free text

3. Other than treating the seizures symptomatically, would you start first-line immunotherapy for presumed autoimmune encephalitis?

- No
- Yes
- Yes, only if autoantibody panel comes back positive (we don't need to ask what type CSF/serum because we have the previous response)
- Free text

If answering no (skip logic):

3b. After two weeks, the autoantibody panel comes back with low-titre positive anti-NMDAR antibodies in both serum and CSF. The patient had two short (< 5 min) focal seizures with alteration of awareness over the past week despite therapy with two appropriately dosed antiepileptic drugs (AEDs). Would you start first-line immunotherapy at this point?

- No, I would make changes to the AEDs and reassess
- Yes, but I would also make changes to the AEDs
- Yes, and I would keep the same AEDs
- Free text

DEMOGRAPHIC QUESTIONS

1. Do you identify as a neuroimmmunologist?

- Yes
- No

2. How many cases of autoimmune encephalitis have you managed in the last 12 months?

- 0
- 1 to 5
- 6 o 10
- 11 to 20
- More than 20

3. What patient population do you treat?

- Adults (18 years and older)
- Children (0 to 18 years)
- Both (adults and children)

4. How many years have you been in practice?

- In Training
- Less than 10 years
- 10 or more years

5. What is your primary work setting?

- Hospital-based (including free-standing, academic, and government)
- Outpatient-based (including neurology clinic, multi-speciality clinic, and private practice)

6. What is your current level of training?

- Attending/Consultant/Faculty Board-certified Physician
- Resident/Fellow
- Physician Assistant (PA)

- Nurse Practitioner (NP)
- 7. Do you currently practice in the United States?
 - Yes
 - No

8a. In what country is your practice located? (If No to 7; Respondent selects from list of all countries)

8b. In what state is your practice located? (If Yes to 7; Respondent selects from list of U.S. States)