Behavior change, depressed mood and catatonia: acute transient psychotic disorder or autoimmune encephalitis?

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Case report

17 y.o girl presented abrupt change in behavior, deteriorating over 4 weeks: confusion, sleeplessness, mutism, depressed mood, feeding disturbance and dyskinetic movement of the orofacial region. Despite slight slowing EEG, first clinical evaluation led to diagnosis of acute transient psychotic disorder treated with risperidone. 48h later she presented general tonico-clonic seizure. 2nd EEG showed extreme delta brushes. Cerebral MRI revealed T2 hyperintensity in the left hippocampus and thalamus. CSF showed no pleicytosis, normal proteinorachy but oligoclonal bands were found. NDMAR antibody were negative in serum but positive in CSF (1/32). No ovarian teratoma was found. Initial treatment included 5 steroid boluses (1g/d) and IV immunoglobulin, followed by rituximab. Secondary to dramatic outbreaks of auto mutilations, she was hospitalized in ICU during several weeks for deep sedation. She recovered slowly but completely except for tiredness.

Discussion

Anti-N-methyl-D-aspartate (NMDA) receptor encephalitis is a severe form of autoimmune encephalitis associated with antibodies against NR1 and NR2 subunits of the NMDA receptor. Initially described in adult women with teratomas, it is now recognized among adult males and children, even without tumor.

The stereotypical clinical course is non-specific flu-like prodrome followed by psychotic stage: bizarre behavior, paranoid thoughts, visual or auditory hallucinations and dyskinesia; some cases of pure psychiatric presentation exist. Brain MRI is either normal (50% cases), or shows non-specific T2 hyperintensity in hippocampus, frontal and insular cortex. EEG is altered with background slowing. CSF abnormalities include lymphocytosis and oligoclonal bands. Diagnosis is confirmed by NMDA receptor antibodies in serum or cerebrospinal fluid. Improvement takes several months. Approximately 80% of patients have substantial or full recovery, with less than 5% mortality. First-step treatment includes steroid bolus and IVIg with eventual tumour resection; second line rituximab and/or cyclophosphamide. Yearly tumour check is advised.

Conclusion

- Think of Anti-NMDA receptor encephalitis if acute and unexplained behavioral changes
- It is a treatable disorder
- It can be an early marker of ovarian teratoma

References

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3. Maarten J Titulaer et al : Treatment and prognostic factors for long-term outcome in patients with anti-NMDA receptor encephalitis: an observational cohort study - Lancet Neurol 2013
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EEG: Extreme delta brushes

Diffusion-weighted MRI : Hyperintensity of the left hippocampus

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