An interesting case of encephalitis

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Autoimmune encephalitis is an emerging clinical entity/medical emergency. Psychiatric symptoms can be a presenting feature of this condition, causing diagnostic confusion. Early and appropriate treatment is essential for complete recovery and to prevent relapse.

Keywords:
anti-*N*-methyl-D-aspartate receptor antibody, autoimmune, encephalitis, ovarian teratoma, psychiatric symptom

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Introduction

Psychiatric presentation of medical illness is not uncommon, but a real diagnostic challenge. Here we discuss the case of a 21-year-old girl presented with excessive fear and agitated behaviour with one episode of tonic posturing, which was a diagnostic dilemma.

Case report

A 21-year-old woman was referred to psychiatry outpatient department with unexplained excessive fear for last 3 weeks. She was also having altered sleep–wake cycle with excessive daytime sleepiness. Occasionally, she showed agitated behaviour and disorientation, during which period she used to repeat the same words and phrases. There was one episode of tonic posturing not associated with loss of consciousness, frothing from mouth, incontinence of bowel/bladder or tongue biting. She had no recent fever or any other significant illness in the immediate past. Neither she nor her family members had any psychiatric illness. Clinical examination was unrevealing except for lack of attention to commands. In view of altered behaviour and seizure-like activity, we made a provisional diagnosis of acute encephalitis. Routine blood examinations including liver function test, renal function test and serum electrolytes were within normal limits. Computed tomography and MRI brain were normal. Cerebrospinal fluid (CSF) showed lymphocytic leucocytosis with normal biochemical parameters. She was started on injection acyclovir intravenously, with a clinical diagnosis of acute viral encephalitis. But there was no significant improvement in her clinical status. So repeat lumbar puncture was done and the sample was sent for autoimmune screening which showed anti-*N*-methyl-D-aspartate receptor (NMDAR) positivity and a clinical diagnosis of anti-NMDAR positive autoimmune encephalitis was made. The patient improved considerably with 5 days of intravenous methyl prednisolone and was switched over to oral prednisolone. We did a malignancy screen for the patient which came as negative. The patient was discharged and kept under follow-up. She improved completely, when reviewed after 15 days.

Discussion

Acute encephalitis is a medical emergency which can be due to infection or autoimmune aetiology. Autoimmunity can be either a part of systemic autoimmune diseases or due to specific antibodies targeting the cellular components of neuronal tissues such as intracellular antigens, synaptic proteins or extracellular domains of cell surface receptors.

Anti-NMDA receptor positive encephalitis, a recently detected clinical entity, was first described in 2007 as a paraneoplastic manifestation of ovarian teratoma. It is characterised by antibodies against the NR1 subunit of the extracellular domain of the NMDAR [1].

Anti-NMDA receptor encephalitis usually affects young women and presents with psychiatric symptoms such as anxiety, sleep disturbance, agitation, mood labiality, hallucination, echolalia, psychosis and catatonia, at the onset, which may be confused initially as primary

psychiatric illness. Following initial psychiatric symptoms, the patient develops seizure, altered level of consciousness, cognitive alteration, dyskinesia, choreoathetoid movements and dystonic posturing. More than 50% of the patients develop autonomic instability in the form of tachycardia, bradycardia, central hypoventilation and hyperthermia which may be confused with neuroleptic malignant syndrome, if the patients were already started on antipsychotic medications [2,3].

Anti-NMDAR positive encephalitis can be paraneoplastic or nonparaneoplastic. About 38% cases are associated with an underlying tumour. Ovarian teratoma is the most common tumour associated with paraneoplastic anti-NMDAR positive encephalitis accounting for about 95% of the cases and other less common tumours associated with this include extra ovarian teratoma, carcinoma of the breast, lung, thymus, testis and pancreas. So a malignancy work up is mandatory for all the patients diagnosed with anti-NMDAR encephalitis [4]. The patient should be kept under follow-up if the initial screening for malignancy is negative.

CSF shows lymphocytic pleocytosis and presence of anti-NMDAR antibody in serum as well as CSF is diagnostic of anti-NMDAR encephalitis [4]. CSF antibody is more specific as serum testing may be false negative in 15% of the cases. Electroencephalogram may show slow disorganised activity or may show seizure activity if the patient is having seizure. MRI may be normal but may show evidence encephalitis, cerebellitis, striatal encephalitis, or brainstem encephalitis [5].

Treatments include surgical removal of the underlying tumour, if any, and immunotherapies including corticosteroids, intravenous immunoglobulin and plasmapheresis. If the patient is not responding to first-line immunosuppressants, second-line immunomodulators such as cyclophosphamide or rituximab is indicated [6]. Psychiatric symptoms are treated with various antipsychotic agents [6].

Patients will recover completely if treated early and appropriately [7]. Relapse can occur in up to 25% patients and the risk for relapse is high in patients treated suboptimally [8].

Awareness about anti-NMDAR encephalitis is essential among physicians as well as among psychiatrists because of initial psychiatric presentation, potential for complete recovery and increasing risk for relapse with delay in diagnosis and treatment.

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Conflicts of interest
There are no conflicts of interest.

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