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Seronegative Autoimmune Encephalitis Presenting as Acute Cerebellitis, A CSOM Complication

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Abstract: ***Neuropsychiatric symptoms are very common in autoimmune encephalopathy. We report a child with seronegative autoimmune encephalitis who presented as acute cerebellitis with Chronic suppurative otitis media (CSOM), initially treated as complication of CSOM however high index of suspicion for autoimmune encephalitis was kept and was managed with intravenous pulse steroid and intravenous immunoglobulins (IVIg).***

Keywords: seronegative autoimmune encephalitis, neuropsychiatric symptoms, pulse steroids

1. Introduction

Autoimmune encephalitis (AE) is a common cause of pediatric encephalopathy. The autoimmune process can be triggered by an infection, vaccine, or malignancy. Neuropsychiatric symptoms are very common in autoimmune encephalopathy and affected children may initially present to psychiatrists. Neurological features include movement disorders, seizures, altered sensorium, and cognitive regression. Hypoventilation and autonomic features can also be associated symptoms. Cerebrospinal fluid examination may suggest inflammatory findings; but are relatively nonspecific. Magnetic resonance imaging (MRI) may also demonstrate abnormalities that provide clues for diagnosis, particularly on fluid-attenuated inversion recovery or T2-weighted images. AE is well responsive to immune therapy, with prompt diagnosis and treatment strongly beneficial. Herein, the authors present a case of AE presented as a neuropsychiatric symptoms mimicking the acute cerebellitis as a complication of CSOM.

2. Case Report

A 5-year-old boy, presented with abnormal behavior during sleep at 4: 00 am with sudden shouting after waking up from bed. Within 2 hours, the child was completely drowsy with decreased interaction with parents and reduced movements. The child had a past history of CSOM like illness since 6 months of age. He had painless purulent ear discharge with decreased hearing from the right ear. He had received medications from various healthcare institutions for his recurrent symptoms. The child had active discharge from right ear during the current admission also. On examination, child was lethargic. He had clasp knife rigidity in all four limbs with brisk reflexes; cerebellar signs were positive in form of scanning speech, truncal ataxia, nystagmus, finger-nose-finger test and shin to heel test positive with no meningeal signs. Possibility of acute cerebellitis, complication of CSOM or autoimmune encephalitis was kept. Complete blood count showed leucocytosis, reactive thrombocytosis with a positive CRP and positive procalcitonin. He was started on intravenous imipenem. Child was afebrile during hospital stay and his cerebrospinal fluid analysis was within normal limits with sterile cultures for CSF and blood. He was started on clonazepam after which there was improvement of movements and sleep. He was continued on valproate, levetiracetam, phenytoin and topiramate. However, he developed seizures during the hospital stay for which phenobarbitone and lacosamide was added and subsequently seizures were controlled.

There was partial improvement in sensorium during the initial hospital stay, but in view of persistent movement disorder, autoimmune encephalitis was considered. He was started on pulse methylprednisolone followed by 2g/kg of IVIg. CSF autoimmune panel and other autoimmune workup came out to be negative. He was managed as a case of sero-negative autoimmune encephalitis and first line immune-modulation was given with a plan to review after 14 days. At the time of discharge the child was able to recognize parents occasionally, accepting feed orally.

3. Discussion

Encephalitis has high incidence and mortality rates worldwide (1) , with a reported mortality rate of 8–18.45% (2, 3) . Our case highlights that the pediatricians need to maintain a high level of suspicion when evaluating children with new-onset seizures, neuropsychiatric symptoms or encephalopathy since autoimmune encephalopathies likely remain underdiagnosed. The importance of considering autoimmune pathogenesis in the differential diagnosis of encephalitis is crucial, as early recognition and treatment may affect the outcome (4) . In our case child presented as acute cerebellitis with a possible underlying bacterial etiology however high suspicion of autoimmune encephalitis was kept and child was given early pulse steroid followed by IVIg which improved the clinical condition of child.

Taken together, all these findings suggest clinicians to give special consideration that the AE are the group of autoantibody-mediated encephalitis that responds well to immunotherapy. The initial diagnostic criteria were very much reliant on antibody testing and response to immunotherapy but these criteria may always not be detectable (5) . Diagnostic challenge occurs when there are no detectable auto-antibodies in serum or CSF.

4. Conflict of interest

None

5. Financial disclosure

None

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