Case Report

Post-vaccinial encephalomyelitis – Probable acute disseminated encephalomyelitis (ADEM) -: A case report

Manish Kumar[1], Sangita Kamath[1], Ashok Sunder[1]

[1]*Department of Internal Medicine, Tata Main Hospital, Tata Steel, Jamshedpur, Jharkhand, India*

Abstract

Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating disease that typically occurs following a viral infection or vaccination. The incidence of ADEM following vaccination has fallen since the introduction of non-neural rabies vaccine and very few cases have been reported due to pure chick embryo derived rabies vaccine (PCERV). Here we are reporting a rare case of probable post vaccinial ADEM.

Keywords: Demyelination, fever, paralysis, vaccine

Introduction

Acute disseminated encephalomyelitis is a demyelinating disease of the central nervous system that typically presents as a monophasic disorder[1-3] with encephalopathy and multifocal neurologic symptoms. However, a relapsing variant (distinct from multiple sclerosis) called multiphasic disseminated encephalomyelitis (MDEM)[4] is also well recognized.

The illness mainly affects children and often occurs a week or so after a viral infection or vaccinations.[5] There seems to be no gender predominance.[5,6,7] Approximately 50–75% of ADEM cases are associated with measles, mumps, rubella, varicella zoster, Epstein-Barr, cytomegalovirus, herpes simplex, hepatitis A, influenza, enterovirus infections, and less than 5% of cases following immunization for rabies, hepatitis B, influenza, Japanese B encephalitis, diphtheria, pertussis, tetanus, measles, mumps, rubella, pneumococcus, polio, smallpox, and varicella suggesting that it is immunologically mediated.[8,9] Additional associated bacterial infections include Leptospira, beta-hemolytic streptococci, and Borrelia burgdorferi.[3,10-12]

The hallmark of ADEM is the presence of widely scattered small foci of perivenular inflammation and demyelination, in contrast to larger confluent demyelinating lesions typical of multiple sclerosis. In the most explosive form of ADEM, acute hemorrhagic leukoencephalitis, the lesions are vasculitic and hemorrhagic, and the clinical course is devastating.[1,13,14] Effective therapy includes high-dose corticosteroids, intravenous immunoglobulins, and plasmapheresis.[1,4] The prognosis is generally favorable almost with full recovery.[1] Here, we describe a case of a 14-year-old child who suffered from probable ADEM two weeks after anti rabies vaccination.

Case Report

A 14-year-old male, was admitted to Tata Main Hospital with complaints of pain abdomen and 6 to 7 episodes of vomiting followed by altered sensorium within 24 hours prior of

Address for correspondence: Dr. Manish Kumar, Department of Internal Medicine, Tata Main Hospital, Tata Steel, Jamshedpur, Jharkhand, India.
E-mail: drmanishkumar2375@gmail.com

Received: 14-08-2019 **Revised:** 16-08-2019 **Accepted:** 05-09-2019

Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/jfmpc.jfmpc_651_19

How to cite this article: Kumar M, Kamath S, Sunder A. Post-vaccinial encephalomyelitis – Probable acute disseminated encephalomyelitis (ADEM) -: A case report. J Family Med Prim Care 2019;8:3425-7.

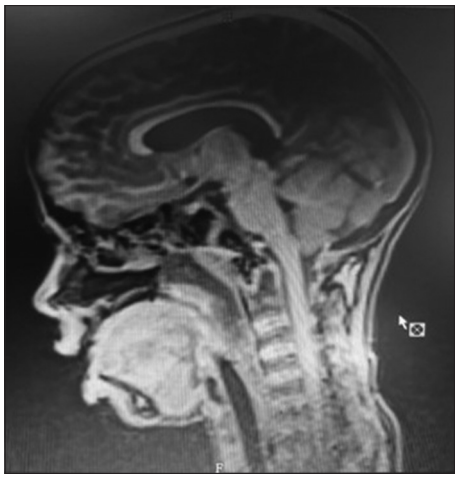

Figure 1: MRI Brain: T1 image sagittal plane shows normal appearance

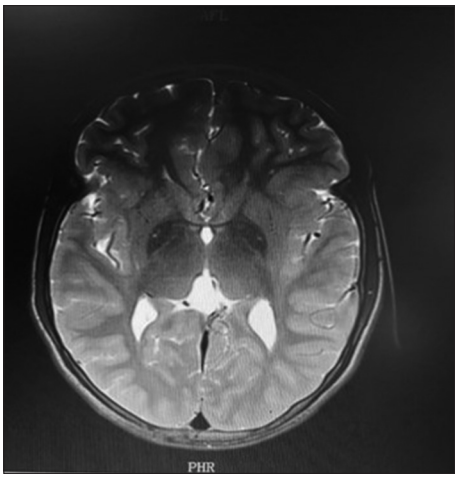

Figure 2: MRI Brain: T2 image transverse plane shows normal appearance

admission. He also had one episode of generalized tonic- clonic convulsion on the way to hospital. He gave a history of dog bite two weeks before his admission and had received three doses of anti-rabies vaccine. He did not have headache, dizziness, and sphincter dysfunction. He had no history of toxic substance abuse, allergy to drugs, surgery, and trauma. He was a full-term baby, born by normal vaginal delivery.

On admission, child did not have pallor, icterus, lymphadenopathy, and was afebrile with Pulse 140/minute, BP- 160/110 mmHg, respiratory rate 14/minuet, near gasping. Examination of central nervous system revealed Glasgow coma score of 4/15 (Eye-1, Verbal-1, and Motor-2), bilaterally dilated pupils with sluggish reaction to light, divergent gaze, hypotonia of all 4 limbs with areflexia and bilaterally extensor plantars. Examination of other systems was normal.

Patient's biochemical investigations revealed fasting blood sugar 102 mg/dl, serum sodium – 148 mmmol/l, serum potassium – 4.2 mmol/l, arterial blood gases (PH – 7.42, PCo2- 38 mmHg, Po2-88 mmHg), serum billirubin -1.0 mg/dl, AST-38U/L, ALT-42U/L, and serum creatinine -0.8 mg/dl. His hemoglobin was 11.8 gm%, TLC-10800/cumm, and platelet count-380000/cumm. Paracheck was negative and malarial parasite was not detected in the peripheral smear. Other investigations including blood and urine cultures were normal. Antinuclear (ANA) antibodies were negative. CSF study showed 5 cells with normal protein and sugar levels (WBC-5 cells, all lymphocytes, sugar-89 mg/dl, protein-37.2 mg/dl, ADA-2.1); Gram stain for micro-organisms was negative. He was non-reactive for HIV l and ll, HCV antibodies, and HBS surface antigen. Chest X ray and USG whole abdomen were normal.

MRI brain and whole spine done on the 2nd day of admission was normal [Figures 1 and 2].

Post admission he was intubated and put on ventilatory support. He was treated with intravenous antibiotics and Methylprednisolone 1 gm per day intravenously for 5 days followed by oral prednisolone 30 mg/day for 5 days. With treatment, his consciousness improved. He was weaned off ventilatory support withdrawn on the 10th day. He was discharged on the 15th day of admission without any neurological deficit. Unfortunately, patient refused to undergo repeat CSF study and MRI brain due to financial constraints.

Case Discussion

Acute disseminated encephalomyelitis is a demyelinating disease of the central nervous system with involvement of the cerebral hemispheres, cerebellum, brainstem, spinal cord, and optic nerves that typically presents as a monophasic disorder, encephalopathy, and multifocal neurologic symptoms. It is caused by immune system dysregulation and triggered by an infectious or other environmental agent in a genetically susceptible host, typically following a recent (1-2 weeks) viral infection or vaccination.

Multiple sclerosis (MS) is an important differential of ADEM initially. MS usually presents as a monosymptomatic disease such as optic neuritis or a sub-acute myelopathy. The optic neuritis in ADEM occurs simultaneously and is bilateral whereas in MS, it is more often unilateral. Fever, meningism and/or psychiatric manifestations which are characteristic of ADEM, are virtually never present in MS.[15]

Our patient had no history of fever or signs of meningism but presented with pain abdomen and recurrent vomiting followed by altered sensorium, had one episode of generalized tonic–clonic convulsion on the way to hospital. He had presented with acute onset of a progressive neurological syndrome affecting multiple cranial nerves, autonomic dysfunction (Pulse 140/minute, BP- 160/110 mmHg), bulbar and pyramidal tracts, following third dose of anti-rabies vaccination.

Points in favor of ADEM:

- History of anti-rabies vaccination
- Rapid deterioration
- Features of encephalopathy

- Multifocal neurologic symptoms (involving higher mental functions, cranial nerves, motor system, autonomic function)
- No other aetiology to explain these events
- Good response to IV methylprednisolone.

Points not in favor of ADEM:
- Normal Cerebrospinal fluid (CSF)
- Normal MRI Brain.

On literature search we found that CSF can be normal in about one-third of such patients and MRI can be normal at the initial presentation abnormalities can develop later (between 5 and 14 days from symptom onset).[11] However we couldn't repeat CSF study and MRI brain because of financial constraints. Also, no case of ADEM with normal MRI brain and CSF study has ever been reported earlier in literature. During his follow up for three months, he never developed any new neurological manifestations and is doing well. The patient's clinical presentation was most likely due to post vaccination Acute Disseminated Encephalomyelitis because no other etiology can explain these events.

Conclusion

A clear-cut latent period between the onset of neurological illness and vaccination, with the typical pattern of diffuse and multifocal involvement of both the central nervous system and peripheral nervous system favors ADEM. CSF and MRI brain may be normal if done early. As methylprednisolone therapy shortens the duration of neurological symptoms and halts further progression, it is important to make an early diagnosis and initiate treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Harrisons Principle of Internal Medicine, 20th edition, chapter 436, page 3200.
2. Nishiyama M, Nagase H, Tomioka K, Tanaka T, Yamaguchi H, Ishida Y, *et al.* Clinical time course of pediatric acute disseminated encephalomyelitis. Brain Dev 2019;41:531-7.
3. Essrani R, Essrani RK, Mehershahi S, Lohana AK, Sudhakaran A. Oculomotor nerve palsy after influenza vaccine in inflammatory bowel disease. Cureus 2018;10:e3759.
4. Bennetto L, Scolding N. Inflammatory/post-infectious encephalomyelitis. J Neurol Neurosurg Psychiatry 2004;75(Suppl 1):i22-8.
5. ZMendoza, Plasencia *et al.* Neurología 2010;25:58-69.
6. Huynh W, Cordato DJ, Kehdi E, Masters LT, Dedousis C. Post-vaccination encephalomyelitis: Literature review and illustrative case. J Clin Neurosci 2008;15:1315-22.
7. G Guliani, Parry (Minneapolis, US). Multiple Sclerosis triggered by purified chicken embryo cell rabies vaccine; 5th Joint triennial congress of the European and American Committees for Treatment and Research in Multiple Sclerosis Amsterdam, The Netherlands 2011;15.30-17.00.
8. Schwarz S, Mohr A, Knauth M, Wildemann B, Storch-Hagenlocher B. Acute disseminated encephalomyelitis: A follow-up study of 40 patients. Neurology 2001;56:1313-8.
9. Kawasaki A, Purvin VA, Tang R. Bilateral anterior ischemic optic neuropathy following influenza vaccination. J Neuroophthalmol 1998;18:56-9.
10. Alves JM, Marques IB, Gil-Gouveia R. [Vaccination controversies: An adult case of post-vaccinal acute disseminated encephalomyelitis]. Acta Med Port 2019;32:81-5.
11. Rossor T, Benetou C, Wright S, Duignan S, Lascelles K, Robinson R, *et al.* Early predictors of epilepsy and subsequent relapse in children with acute disseminated encephalomyelitis. Mult Scler 2019:1352458518823486.
12. Galetta KM, Bhattacharyya S. Multiple sclerosis and autoimmune neurology of the central nervous system. Med Clin North Am 2019;103:325-36.
13. Kashyap R, Jaret P, Mahajan S, Thakur J, Sharma R, Patial RK, *et al.* Post-vaccinal encephalitis after semple type anti-rabies vaccine. JIACM 2004;5:281-3.
14. Harloff A, Rauer S, Hofer M, Klisch J, Els T. Fulminant acute disseminated encephalomyelitis mimicking acute bacterial meningoencephalitis. Eur J Neurol 2005;12:67-9.
15. Kulkarni V, Nadgir D, Tapiawala S, Malabari A, Kalgikar A, Kela R, *et al.* Biphasic demyelination of the nervous system. Neurol India 2004;52:106-8.