

A CASE REPORT ON ACUTE DISSEMINATED ENCEPHALOMYELITIS

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ABSTRACT

Acute disseminated encephalomyelitis (ADEM) is a rare, inflammatory, demyelinating disease of the central nervous system. It predominantly affects children, but it can occur in adults too. We present a case of a 6-year-old child who came to the hospital with complaints of recurrent abdominal pain, vomiting, stiffening of upper limbs, and flexed posture of the left upper limb. His lab investigations revealed leukocytosis and elevated CRP. The CSF examination revealed pleocytosis and elevated protein. The MRI showed hyperintense lesions on T2 and FLAIR sequences over the cortical and subcortical white matter. Hence, a diagnosis of ADEM was made, and the condition was managed with corticosteroids, intravenous immunoglobulins, and other supportive measures.

KEYWORDS: Acute disseminated encephalomyelitis, Demyelinating disease, Leukocytosis, Upper limbs stiffening, Hyperintense lesions, Autoimmune disease.

INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is an autoimmune, rare neurological condition in which widespread inflammation of the central nervous system (brain and spinal cord) causes damage to the myelin sheath (demyelination). Demyelination affects the ability of the nerves to transmit information and causes a wide range of neurological symptoms.^[1] This condition usually occurs in response to a preceding infection or immunization.^[2] ADEM can occur at any age, but children are most commonly affected.^[3]

CASE PRESENTATION

A 6-year-old boy was brought into the hospital with complaints of recurrent abdominal pain, vomiting, stiffening of upper limbs, and flexed posture of left upper limb for 3 days. The patient was normal three days back and developed a

flexed posture of the left upper limb. His history shows that he had a high-grade intermittent fever ten days back, which was relieved with medications. The patient was admitted to the intensive care unit and on examination, the patient was found to be conscious, alert, and febrile with stable vital signs. Hypertonia was higher in the left upper limb than the right upper limb with the presence of tremors.

Table 1: Laboratory investigations.

| PARAMETERS | VALUE |
|------------|------------------------------|
| Hemoglobin | 13.7 g/dL |
| WBC | 19,400 cells/mm ³ |
| RBC | 3.86 million/mm ³ |
| Platelets | 2.1 lakhs/mm ³ |
| CRP | 110.11 mg/L |
| HIV | Negative |
| HBsAg | Negative |

Laboratory investigations revealed an elevated WBC count of 19,400 cells/mm³ and an increased CRP level of 110.11 mg/L. Serology for HIV, and HBsAg were negative.

A lumbar puncture was done, and the CSF was examined which showed elevation in the white blood cell count and increased protein concentration.

Table 2: CSF examination.

| PARAMETERS | VALUE |
|----------------------|-----------------|
| Colour | Colourless |
| Appearance | Slightly turbid |
| TLC | 280 cells |
| Biochemical analysis | |
| Glucose | 64 mg/dL |
| Protein | 104 mg/dL |

MRI-Brain revealed hyperintense lesions on T2 and FLAIR sequences over the cortical and subcortical white matter.

The patient was initiated on IV Ceftriaxone (1 g BD), IV Methylprednisolone (250 mg OD), and supportive care measures. On the third day, the patient experienced a seizure episode and was thus put on IV Levetiracetam (250 mg BD). On the fifth day, the patient was administered intravenous immunoglobulin (5 g OD). Initially admitted to the ICU, the patient was then transferred to the medical ward on the eighth day. After recovering, the patient was discharged from the hospital. Upon discharge, he was prescribed with Tab Brivaracetam (25 mg, Half tablet BD), Syp Prednisolone (5 ml OD), and other supportive treatments.

DISCUSSION

ADEM is a rare, autoimmune neurological disorder that causes inflammation in the central nervous system leading to damage of the myelin sheath.^[4] Studies estimate the annual incidence of ADEM to be 0.07 to 0.4 per 100,000 population per year.^[5] The exact cause of ADEM is unknown, but it may be associated with an immune response towards post-infection or immunization. ADEM may occur following bacterial or viral infection (like measles, mumps, rubella, influenza, Varicella zoster, Epstein-Barr virus, Cytomegalovirus, Herpes simplex virus, Hepatitis A & C virus, Dengue virus, Borrelia burgdorferi, Chlamydia, Legionella, Mycoplasma pneumoniae, Rickettsia rickettsii, Streptococci, Plasmodium vivax).^[6,7] ADEM may also occur following vaccination, but this is very rare and noted in only <5% of all ADEM cases.^[1]

The exact mechanism of ADEM is not entirely understood, theories include either a cell-mediated response or antibodies produced in response to a trigger that targets the myelin protein, leading to demyelination. Another theory is the increased vascular permeability and congestion in CNS triggered by an inflammation and circulating immune complexes following vaccination or infection. Then demyelination, gliosis, and necrosis happen as a result of the inflammatory cascade involving peri-venous hemorrhage, edema, and infiltration of inflammatory cells.^[2,8]

The laboratory workup for ADEM shows raised inflammatory markers, and CSF studies show elevated protein and lymphocytic pleocytosis. Bilateral and asymmetrical lesions on MRI in the white matter are the hallmarks of ADEM. The lesions are hyperintense on T2 weighted and FLAIR sequences.^[9]

Immunomodulatory and supportive therapies are used in the treatment of ADEM. Corticosteroids, intravenous immunoglobulins, and plasma exchange are the potential treatment options. Intravenous Methylprednisolone is the first line of therapy (10-30 mg/kg/day, maximum 1g/day) for 3-5 days followed by oral corticosteroids continued with gradual tapering over 6 weeks to reduce the risk of relapses. The second line of treatment includes intravenous immunoglobulins (IVIg) (0.4 g/kg/day for 2-5 days) and plasma exchange. This treatment is highly effective in steroid non-responders.^[10]

The prognosis of ADEM is good among pediatric patients with complete recovery in about 80% of the cases. 10% of the children initially diagnosed with ADEM may experience another ADEM attack, typically within 2-8 years of the first attack. However, 15% of the adults may develop multiphasic disseminated encephalomyelitis, a recurrence of the disease.^[8]

It is evident that ADEM encompasses a significant portion of the spectrum of CNS inflammatory demyelinating diseases. To identify the pathogenic and immunological features of the illness, more research is required.^[8]

CONCLUSION

This case study reveals the presentation of acute disseminated encephalomyelitis (ADEM), a rare neurological condition observed in a 6-year-old child. The patient's condition was initially managed with corticosteroids, and when symptoms did not resolve, intravenous immunoglobulins were added to the treatment plan, leading to the patient's recovery.

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Conflict of interest

There is no conflict of interest.

REFERENCES

1. Acute Disseminated Encephalomyelitis - Symptoms, Causes, Treatment | NORD [Internet]. [cited 2024 Aug 22]. Available from: <https://rarediseases.org/rare-diseases/acute-disseminated-encephalomyelitis/>
2. Anilkumar AC, Foris LA, Tadi P. Acute Disseminated Encephalomyelitis. In: Stat Pearls [Internet]. Treasure Island (FL): Stat Pearls Publishing; 2024 [cited 2024 Aug 22]. Available from: <http://www.ncbi.nlm.nih.gov/books/NBK430934/>

3. Acute Disseminated Encephalomyelitis (ADEM) | Boston Children's Hospital [Internet]. [cited 2024 Aug 22]. Available from: <https://www.childrenshospital.org/conditions/acute-disseminated-encephalomyelitis-adem>
4. Acute Disseminated Encephalomyelitis (ADEM): Symptoms [Internet]. [cited 2024 Aug 22]. Available from: <https://my.clevelandclinic.org/health/diseases/14266-acute-disseminated-encephalomyelitis-adem>
5. Balamurugesan K, Ponprabha R, Davis P. An interesting case of acute disseminated encephalomyelitis following E. coli infection. *J Family Med Prim Care*, 2019 Jan; 8(1): 292–5.
6. Savitha M, Sudagar Singh RB, Damodharan J, Sivaprakash V. POSSIBLE ADEM - A CASE REPORT [Internet]. Edu.in. [cited 2024 Aug 22]. Available from: https://www.sriramachandra.edu.in/university/pdf/research/journals/nov_2014/possible_adem.pdf
7. Balamurugesan K, Ponprabha R, Davis P. An interesting case of acute disseminated encephalomyelitis following E. coli infection. *J Family Med Prim Care*, 2019 Jan; 8(1): 292–5.
8. Almaghrabi N, Saab A. Adult-onset acute disseminated encephalomyelitis: A case report. *Radiol Case Rep*, 2021 Jul 2; 16(9): 2469–73.
9. Langley L, Zeicu C, Whitton L, Pauls M. Acute disseminated encephalomyelitis (ADEM) associated with COVID-19. *BMJ Case Rep.*, 2020 Dec 13; 13(12): e239597.
10. Acute Disseminated Encephalomyelitis: Case Report and Brief Review - PMC [Internet]. [cited 2024 Aug 22]. Available from: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4311361/>