CASE REPORT

A Fatal Case of Acute Disseminated Encephalomyelitis: A Diagnosis to Ponder in Pandemic

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Abstract

A 40-year-old woman known hypertensive presented with progressive ascending paralysis. MRI T2W and FLAIR screening of the brain demonstrated swelling with altered signal in the visual cervical cord, medulla, and another juxtacortical lesion in the right temporal lobe with possibility of a demyelinating etiology. CSF testing did not identify a direct cerebral infection. High-dose steroids followed by a course of IVIG was administered but with no significant response. In these pandemic times, the patients who present with altered mentation and polyfocal neurological deficits and background history of recent COVID-19 infection or recipient of SARS-CoV-2 vaccine the diagnosis of acute disseminated encephalomyelitis (ADEM) should be considered likely.

Keywords: Acute disseminated encephalomyelitis, COVID-19, Vaccine.

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Introduction

Acute disseminated encephalomyelitis (ADEM) is an immune-mediated demyelinating disorder involving the brain and spinal cord.[1] Coronavirus disease-2019 (COVID-19) is a disease with a significantly broad spectrum of presentation and clinical syndromes. This novel virus has been associated with acute respiratory distress syndrome (ARDS), thromboembolic syndrome, severe metabolic syndromes, severe acute tubular necrosis, electrolyte abnormalities, neurologic syndromes, and cardiac events. Neurological involvement is commonly encountered in critically ill patients and has been widely reported during the current COVID-19 pandemic likely due to inherited viral infection-related immune activation.[2,3] Vaccines also have been implicated as one of the etiological agents for causation of ADEM although rare.[4] Postvaccine ADEM has been associated with various vaccines such as rabies, smallpox, measles, rubella, polio, and influenza vaccine.[5] Though after reviewing the literature, we found case reports of ADEM following COVID-19 virus but, on the contrary, only two case reports of ADEM following SARS-CoV-2 vaccine.[6,7]

Case Description

Forty-year-old female with past history of hypertension on regular treatment with tablet amlodipine received the first dose of ChAd0x1 nCoV-19 corona virus vaccine (recombinant, replication-deficient chimpanzee adenovirus vector encoding the SARS-CoV-2: spike glycoprotein, Serum institute of India private limited, AstraZeneca, India). After 14 days of vaccination, she was hospitalized with complains of acute onset of severe back pain without any radiation for which she received intravenous analgesics that provided a temporary relief. On further probing, she has history of nonspecific myalgia 1 week before vaccination which subsided on its own. Next morning her symptoms progressed to bilateral lower limb weakness with belt fastening sensation around mid-trunk, five fingers above umbilicus. Magnetic resonance imaging (MRI) of whole spine screening was done the next day to investigate the cause of ascending weakness which revealed acute anterior horn hypoperfusion/ischemia at T3–T11 level with hyperintense signals at T9–T11 cord likely cytotoxic edema or hypoperfusion. They detected owls eye sign suggestive of anterior horn ischemia extending over six thoracic segments on postgadolinium contrast-enhanced images with restricted diffusion in some of these segments on DWI. On central nervous system examination, she was dull but vocalizing, no diplopia, no difficulty in swallowing or retention of secretion but had reduced sensation to touch and pain in lower limbs extending till xiphisternum. Within next 24 hours, motor examination revealed 0/5 power in all four limbs as per medical research council grading and bilateral deep tendon reflexes were exaggerated with extensor plantar response. Diagnostic spinal digital subtraction angiography (DSA) revealed grossly normal study. Next day, in view of altered sensorium, respiratory distress, and paradoxical breathing, patient was intubated and put on mechanical ventilation. MRI of brain was done that revealed a swelling with altered signal in the visualized cervical cord-medulla and another juxtacortical lesion in the right temporal lobe on T2 and FLAIR scan suggestive of demyelinating pathology likely ADEM (Fig. 1). In particular, cerebrospinal fluid (CSF) was acellular with normal protein content (48 mg/dL) with no oligoclonal bands. Gram stain and culture, fungal smear, gene Xpert, cryptococcal antigen, TB-gene Xpert, AFB and CSF-EBV PCR, CSF CMV PCR, CSF VZV, and CSF VDRL all were negative. She was started on methylprednisolone 1 g/day after consultation with neurologist. To rule autoimmune encephalitis, NMDA-NR1,

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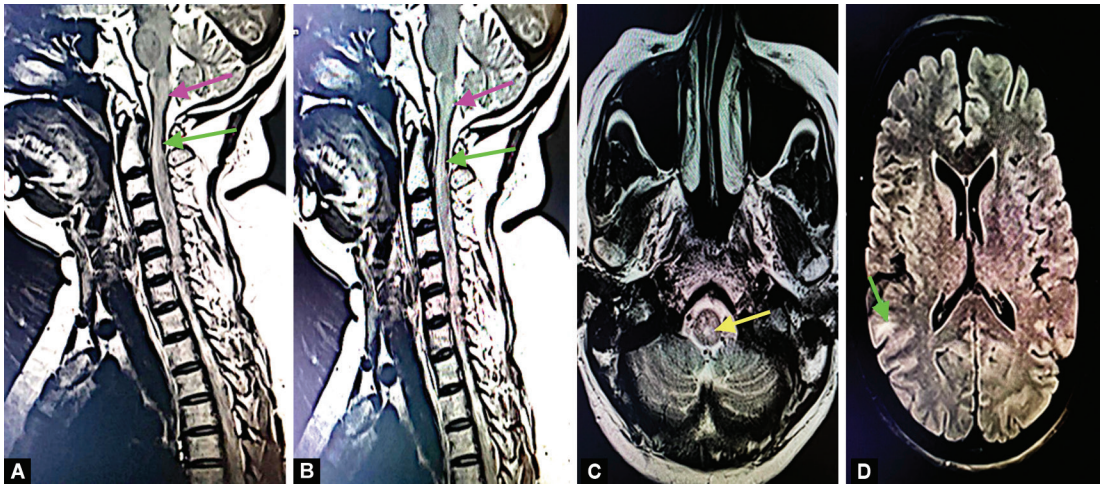

Figs 1A to D: MRI findings suggestive of ADEM

AMPA-GluR1, AMPA-GluR2, GABA-B receptor antibody, LGi-1 antibody, CASPR2 antibody in CSF, and serum were sent but were negative. Anti-double-stranded DNA and extractable nuclear antigen (ENA) panel and antimyelin oligodendrocyte (MOG) antibodies were also negative in serum. Her C3 and C4 levels were normal. It was differentiated from multiple sclerosis on the basis of widespread cord and brain involvement and also first episode of this kind. CSF COVID-19 IgG level was 4.87, while serum COVID-19 IgG was 17.5. Her nasal swab COVID-19 RT-PCR was negative. Patient had features suggestive of dysautonomia with frequent variations in heart rate, blood pressure, and temperature. Treatment was upgraded to intravenous immunoglobulin of 0.4g/kg/day. Unfortunately, patient continued to deteriorate in spite of ventilatory and vasopressor support. Despite the best resuscitative efforts, the patient could not be revived and passed away within a week of admission with such a rapid and fulminant course.

Discussion

Our patient met the diagnostic criteria of ADEM defined by level II of diagnostic certainty by Sejvar et al. and the Brighton Collaboration Encephalitis Working Group, and alternative diagnoses such as Guillian–Barre syndrome, multiple sclerosis, transverse myelitis, and infectious and autoimmune encephalitis were all excluded in our patient.[8] ADEM following SARS-CoV-2 infection has been well documented though there has been sparse data following SARS-CoV-2 vaccine. Although past viral or COVID-19 infection cannot be ruled out due to nonspecific myalgia in past one week before vaccination. The two case reports mentioning about the occurrence of ADEM post-COVID-19 vaccine were both inactivated vaccines in contrary to ours which was recombinant one. The presentation was way different in both reports varying from fever, muscle stiffness, to seizures though, in our case, it was ascending paralysis and, in later stages, it affected the level of consciousness.[7,8] In both the cases referred above, the condition improved dramatically after steroids but this was not observed in our case in spite of intravenous immunoglobulin therapy too. Such a fulminant presentation of ADEM similar to ours has not been reported before in literature.

This condition though extremely rare should be kept in mind when patients present with ascending paralysis or other neurological features especially following recent COVID-19 infection and SARS-CoV-2 vaccination.

Learning Point

Clinicians dealing with adults having neurological involvement with history of COVID-19 infection or respective vaccine recipient, some times it becomes difficult to implicate the cause of multifocal neurological symptoms and thus a high index of suspicion for ADEM should be kept as differential. Early detailed imaging along with CSF examination and aggressive immunosuppressive therapy might be the key.

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