Case Report

Acute disseminated encephalomyelitis in a SARS-CoV2 seropositive child

Pankaj Sahu[1], Ritu[2]

From [1]Senior Resident, [2]Medical Officer, Department of Pediatrics, Dr. Baba Saheb Ambedkar Medical College and Hospital, Rohini, New Delhi, India

ABSTRACT

Children with severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection have various clinical presentations. Neurological manifestations of SARS-CoV-2 have been described in adults but such presentations in children are rare. The different neurological manifestations associated with SARS-CoV-2 are headache, seizures, anosmia, ischemic stroke, encephalopathy, transverse myelitis, and Guillain-Barre Syndrome. Here, we describe a case of SARS-CoV-2 seropositive child presenting as acute disseminated encephalomyelitis.

Key words: Acute disseminated encephalomyelitis, Children, Coronavirus disease 2019, Neurological involvement

Severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection has varied presentations in the pediatric age group. Although most of the infections seen in children are mild, few children suffer severe systemic illness. The different neurological manifestations reported in adults with SARS-CoV-2 include headache, seizures, anosmia, ischemic stroke, encephalopathy, transverse myelitis, and Guillain-Barre Syndrome [1].

Neurological features have been described in about one-third cases of multisystem inflammatory syndrome in children temporally related to SARS-CoV92 [2,3]. In addition, isolated neurological involvement has also been reported in children [4,5]. Here, we describe a case of SARS-CoV-2 seropositive child presenting as acute disseminated encephalomyelitis (ADEM).

CASE REPORT

A previously well 5-year-old boy presented with low grade fever for 7 days, multiple episodes of generalized seizures for 1 day along with sudden onset bilateral lower limb weakness and urinary retention. He did not have cough, headache, vomiting, diarrhea, or rash.

On examination, child was afebrile but drowsy with a heart rate of 104 beats/min, respiratory rate - 22/min, BP - 130/90 mm-Hg, and oxygen saturation of 98% on room air. Neurological examination revealed Glasgow Coma Scale score of 11/15 with normal cranial nerve and fundus examination; however, signs of meningeal irritation (positive Kernig's sign, and neck rigidity) were present. Motor system examination revealed 3/5 power (Medical research council grade) with normal reflexes in the upper limbs and 1/5 power with hyporeflexia in both legs. He also had truncal weakness and bladder involvement but no diaphragmatic weakness. His superficial abdominal reflexes were absent as well and plantar reflexes were bilaterally extensor. Rest of the systemic examination was normal.

Blood investigations showed leukocytosis (Total Leukocyte Count-12.9 × 10^9 cells/L, 73% polymorphs) with unremarkable liver and renal function tests and normal electrolytes. He had a positive C-reactive protein (1.2 mg/dL) but normal ESR (15 mm/h), triglycerides (127 mg/dL), D-dimer (101.8 ng/mL), and ferritin levels (210 ng/mL). Cerebrospinal fluid (CSF) examination revealed 3 cells/mm^3 (100% lymphocytes), protein-52 mg/dL, and sugar 61 mg/dL. CSF analysis was negative for Japanese encephalitis virus, herpes simplex virus, and *Mycobacterium tuberculosis*.

Nasopharyngeal swab reverse transcriptase polymerase chain reaction (RT-PCR) was negative for SARS-CoV-2. However, the serology for SARS-CoV2 (total immunoglobulin IgG and IgM) was positive [69 Cutoff index (COI), COI>1:positive] by electrochemiluminescence assay. Magnetic resonance imaging (MRI) of brain done on day 4 of admission showed multifocal T2/FLAIR hyperintensities in right frontal lobe, bilateral periventricular regions, and left sub cortical white matter (Fig. 1). MRI of the spine revealed longitudinal, patchy T2 hyperintense lesions in dorsal column extending from D1 to D12 vertebra level along with cord expansion (Fig. 2). Based on the neuroimaging, a diagnosis of ADEM with longitudinally extensive myelitis was made.

Access this article online

Received - 15 May 2021
Initial Review - 03 June 2021
Accepted - 24 June 2021

DOI: 10.32677/IJCH.2021.v08.i07.004

Quick Response code

Correspondence to: Dr. Ritu, Department of Pediatrics, Dr Baba Saheb Ambedkar Medical College and Hospital, Rohini, New Delhi, India. E-mail: season.ritu9@gmail.com

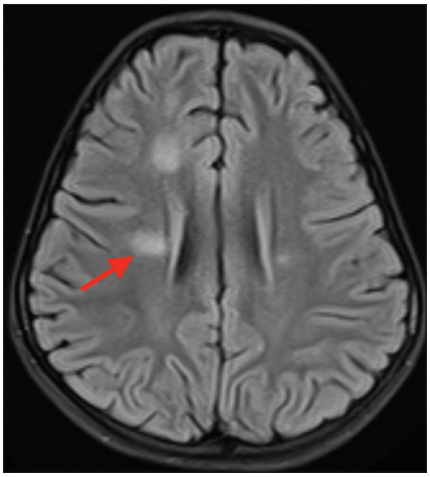

Figure 1: Axial FLAIR magnetic resonance imaging brain showing multiple hyperintense lesions in bilateral periventricular regions (arrow)

Child was initially managed with broad spectrum antibiotics, acyclovir, and antiepileptic drugs. Following confirmation of diagnosis of ADEM on neuroimaging, pulse dose of methyl prednisolone (30 mg/kg/day) was administered for 5 days. There was significant improvement in the muscle strength in both lower limbs by day 5 of pulse therapy and child was transitioned to oral steroids with slow tapering over the next 6 weeks. He was discharged on the 10th day once he improved clinically. On follow-up, the child is well and there is no residual neurologic deficit.

DISCUSSION

ADEM is an immune mediated, multifocal demyelinating illness precipitated following viral infections or immunization [6]. It has been reported in 0.2–14% of adult patients with SARS-CoV-2 infection [1,7]. Few anecdotal reports of ADEM in children with coronavirus disease 2019 (COVID-19) infection have been described as well [4,8].

SARS-CoV-2 associated neurological complications are postulated to be due to either virus mediated neuronal injury, hyperinflammation, and immune mediated (post-infectious) or secondary to a hypercoagulable, thrombogenic state. SARS-CoV-2 belongs to beta *Coronaviridae* that have neuro-invasive propensity [9]. The possible routes for neuro-invasion described are retrograde transmission through the olfactory nerve or hematogenous spread [10]. The cytokine storm following viral invasion releases potent chemotactic mediators (IL-6, 8, monocyte chemotactic protein) that breach the blood brain barrier resulting in intracranial inflammation [1]. Moreover, the receptor for the cellular entry of SARS-CoV-2, that is, Angiotensin converting enzyme 2, is expressed in microglia and neurons of brainstem, neocortex, and spinal cord [11].

The neurological symptoms in COVID-19 infection can occur either acutely (seizures and headache) or have a delayed post-infectious presentation (ADEM). Due to its varied pathophysiological mechanisms, SARS-CoV-2 may or may not

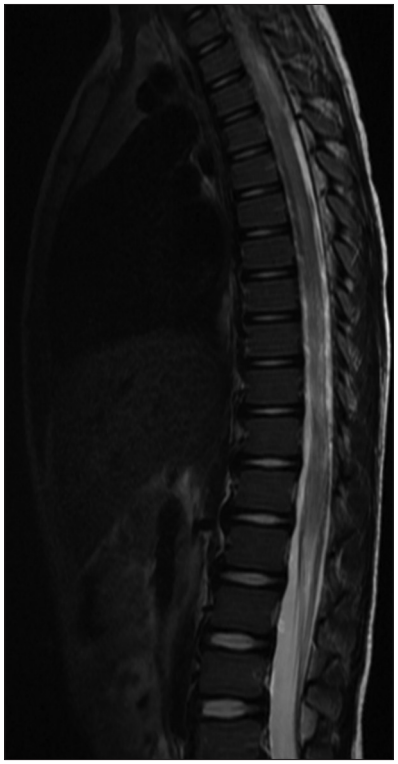

Figure 2: Sagittal view of magnetic resonance imaging dorsal spine showing T2 hyperintensities extending from D1-D12 vertebrae with cord edema

be detected in the CSF [5,8]. The serological tests for SARS-CoV-2 (IgM, IgG, and total antibody) can be used for diagnosing suspected patients who present later on in the illness course and have negative nasopharyngeal RT-PCR. These tests carry high sensitivity (89% for IgM, 95% for IgG, and 98% for total antibody) and high specificity (98% for IgM, 99% for IgG, and 100% for total antibody) after the 2nd week of illness and are helpful for diagnosing past infection, post-COVID-19 sequelae or complications [12].

The typical MRI features of ADEM are patchy, asymmetric, ill-defined T2 hyperintense lesions seen in subcortical white matter, basal ganglia, thalami, and brainstem. Spinal cord lesions are seen in 11–28% patients as longitudinally extensive intramedullary lesions [13]. In a multicentric review of neuroimaging features of children with COVID-19, ADEM like features were seen in 42% and myelitis was seen in 21% cases [14]. The CSF findings include mild lymphocytic pleocytosis with increased protein and normal glucose [13].

Initial therapy with high dose (20–30 mg/kg/day) steroids (methylprednisolone) for 3–5 days, followed by tapering over 4–6 weeks has a complete response in 50–80% patients as was in our case [15]. Intravenous immunoglobulin and plasmapheresis are considered in severe or steroid unresponsive cases [6].

CONCLUSION

Through this case report, we aim to highlight an unusual post-infectious neurological complication of SARS-CoV-2 infection in children and its successful management.

REFERENCES

1. Correia AO, Feitosa PW, de Sousa Moreira JL, Nogueira SÁ, Fonseca RB, Nobre ME. Neurological manifestations of COVID-19 and other coronaviruses: A systematic review. Neurol Psychiatry Brain Res 2020;37:27-32.
2. Chen TH. Neurological involvement associated with COVID-19 infection in children. J Neurol Sci 2020;418:117096.
3. Dufort EM, Koumans EH, Chow EJ, Rosenthal EM, Muse A, Rowlands J, *et al*. Multisystem inflammatory syndrome in children in New York state. N Engl J Med 2020;383:347-58.
4. Mehra B, Aggarwal V, Kumar P, Kundal M, Gupta D, Kumar A, *et al*. COVID-19-associated severe multisystem inflammatory syndrome in children with encephalopathy and neuropathy in an adolescent girl with the successful outcome: An unusual presentation. Indian J Crit Care Med 2020;24:1276-8.
5. Pandey M. Acute meningoencephalitis in a child secondary to SARS-CoV-2 virus. Indian Pediatr 2021;58:183.
6. Pohl D, Alper G, Van Haren K, Kornberg AJ, Lucchinetti CF, Tenembaum S, *et al*. Acute disseminated encephalomyelitis: Updates on an inflammatory CNS syndrome. Neurology 2016;87 9 Suppl 2:S38-45.
7. Paterson RW, Brown RL, Benjamin L, Nortley R, Wiethoff S, Bharucha T, *et al*. The emerging spectrum of COVID-19 neurology: Clinical, radiological and laboratory findings. Brain 2020;143:3104-20.
8. de Miranda Henriques-Souza AM, de Melo AC, Madeiro BD, Freitas LF, Rocha-Filho PA, Gonçalves FG. Acute disseminated encephalomyelitis in a COVID-19 pediatric patient. Neuroradiology 2021;63:141-5.
9. Steardo L, Steardo L Jr., Zorec R, Verkhratsky A. Neuroinfection may contribute to pathophysiology and clinical manifestations of COVID-19. Acta Physiol 2020;229:e13473.
10. Lahiri D, Mondal R, Deb S, Bandyopadhyay D, Shome G, Sarkar S, *et al*. Neuroinvasive potential of a primary respiratory pathogen SARS-CoV2: Summarizing the evidences. Diabetes Metab Syndr Clin Res Rev 2020;14:1053-60.
11. Nemoto W, Yamagata R, Nakagawasai O, Nakagawa K, Hung WY, Fujita M, *et al*. Effect of spinal angiotensin-converting enzyme 2 activation on the formalin-induced nociceptive response in mice. Eur J Pharmacol 2020;872:172950.
12. Hanson KE, Caliendo AM, Arias CA, Englund JA, Hayden MK, Lee MJ, *et al*. Infectious Diseases Society of America Guidelines on the Diagnosis of COVID-19: Serologic Testing. Clin Infect Dis 2020;2020:1343.
13. Tenembaum SN. Acute disseminated encephalomyelitis. Handb Clin Neurol 2013;112:1253-62.
14. Lindan CE, Mankad K, Ram D, Kociolek LK, Silvera VM, Boddaert N, *et al*. Neuroimaging manifestations in children with SARS-CoV-2 infection: A multinational, multicentre collaborative study. Lancet Child Adolesc Health 2020;5:167-77.
15. Alexander M, Murthy JM. Acute disseminated encephalomyelitis: Treatment guidelines. Ann Indian Acad Neurol 2011;14 Suppl 1:S60.

Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Sahu P, Ritu. Acute disseminated encephalomyelitis in a SARS-CoV2 seropositive child. Indian J Child Health. 2021; 8(7):249-251.