

Rhombencephalitis Associated with Sars-Cov-2 Infection in a Girl

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Abstract

Since the beginning of the SARS-CoV-2 pandemic, different neurological complications have been reported during acute infection or convalescence, affecting both adults and children. The following describes the first case of rhombencephalitis in a child with SARS-CoV-2 virus infection.

Keywords: SARS-CoV-2; Rhombencephalitis; Paediatrics

Introduction

Rhombencephalitis is a rare condition in paediatrics [1]. It is caused mainly by infectious agents such as enterovirus 71, herpes virus and *L. monocytogenes* [2]. Diverse neuropathological conditions have related to SARS-CoV-2 infections, which can be associated with multi-systemic involvement or presented as isolated syndromes. The pathophysiological mechanisms can be immune-mediated, postinfectious, or neuro-invasive when the virus is isolated in CSF [3].

Case Description

A previously healthy nine-year-old girl from a rural area (Colombia) presented to the emergency department with a history of fever, odynophagia, diarrhoea and vomiting; symptoms that gradually improved by the 5th day. On day six, the patient complained of dry cough, progressive dyspnea and muscular weakness in the lower limbs, followed by ataxia and impaired speech. On admission, her breath pattern deteriorated and progressed to respiratory failure requiring invasive mechanical ventilation.

A chest X-ray revealed diffuse pulmonary alveolar infiltrates. Arterial blood gas revealed respiratory acidosis with moderate hypoxia. A complete blood count test identified lymphopenia. A metabolic panel evidenced elevated transaminase levels, renal impairment, and an increase of acute-phase reactants including D dimer (2.5 mg/dl) and ferritin (913 ng/mL). Blood cultures were negative, and a serological test ruled out dengue fever.

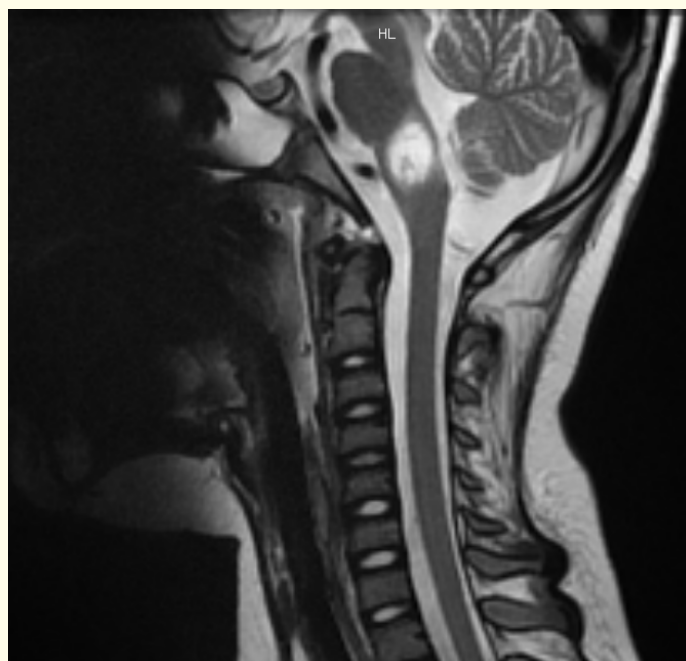
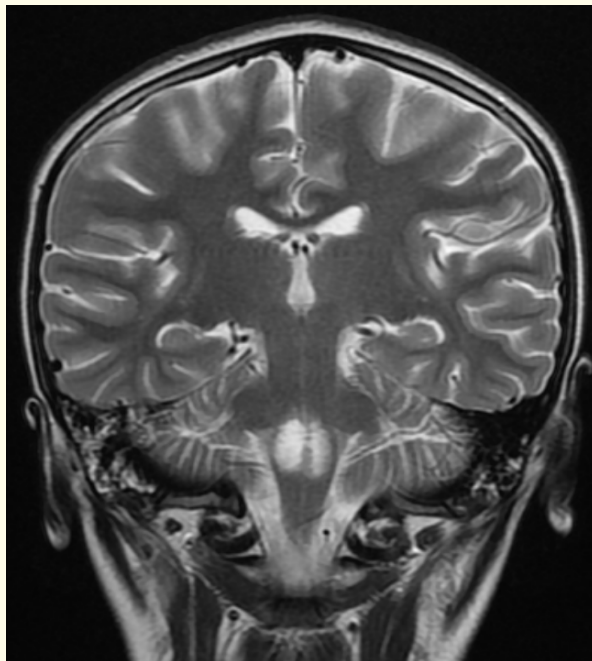
The echocardiogram showed moderate biventricular dysfunction with ejection fraction LVEF 47% and distal ectasia of the left coronary artery. A nasopharyngeal swab RT-PCR test was positive for SARS-CoV-2 infection on day 11. Although pneumonia improved, two extubation attempts failed.

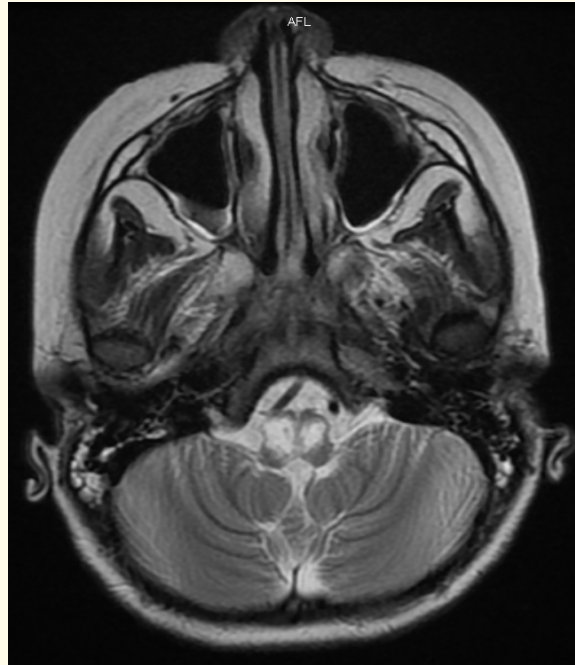
On day 24 sedative medication was reduced, which allowed identifying right hemiparesis. An electromyography and nerve conduction study of the four extremities was performed. The result showed a low grade sensitive and motor myelinic multiple mononeuropathy without signs of denervation, which ruled out Guillain Barré syndrome.

On day 31, under mechanical ventilation, the patient was alert and showed spontaneous facial mimicry. A neurological examination showed flaccid quadriplegia without areflexia, with muscle strength score in neck 1/5, right upper limb 2/5, left upper limb 1/5 and lower limbs 3/5. She presented a normal pupillary reaction to light, normal eye movements, no sensitive level or sphincter dysfunction.

Subsequently a lumbar puncture was performed, showing a clear, colourless liquid, leukocytes 5/mm³, PMN 20%, lymphocytes 80%, glucose 79 mg/dL, and protein 14 mg/dl. The cerebrospinal fluid (CSF) culture was negative for common germs as well as the serology for SARS-Cov-2 (by RT-PCR). A second nasopharyngeal swab RT-PCR SARS-CoV-2 tested negative.

On day 36, an MRI of the brain and spine revealed two hyperintense lesions adjacent to the medulla oblongata on T2 WI and FLAIR sequences, with a diameter of 15 x 10 mm. Areas of contrast enhancement or diffusion restriction were not identified. Another brain and spinal cord injuries were ruled out (Figure 1-3). The diagnoses of rhombencephalitis and Kawasaki-like syndrome linked to COVID 19 were proposed. Consequently, the patient received a single dose of human immunoglobulin at 2 gr/kg and ASA at anti-inflammatory doses. Finally, after three weeks, the patient had a favourable evolution, her muscular strength improved and mechanical ventilation was weaned. At hospital discharge a slight distal tremor of low amplitude remained.





Discussion

The neurovirulence of SARS-CoV-2 can explain the high prevalence of neurological complications, which have been reported up to 36.4% in hospitalized patients with serious and critical condition. Findings predominant among those who develop an excessive immune-mediated inflammatory response, for instance, high levels of CRP, ferritin, and D-dimer [4].

Since the beginning of the SARS-CoV-2 pandemic, two cases of rhombencephalitis have been described in adults associated with systemic infection, none with virus isolation in CSF. The first case, a 40-year-old man whose brain MRI revealed an acute inflammatory lesion in the right inferior cerebellar peduncle and spontaneous remission of neurological symptoms [5]. The second case, a 65-year-old woman whose brain MRI was normal, so that, diagnosis of post-viral encephalitis of the brainstem was considered when improving with steroids [6].

We illustrated a case of rhombencephalitis associated with systemic infection by SARS-CoV2 in a school-age girl without comorbidities, whose clinical course was characterised by early multisystemic disturbance, who had a good clinical response to immunoglobulin treatment.

Although RT-PCR test for SARS-CoV-2 was negative in the CSF, our diagnosis was based on MRI features and CSF findings with lymphocytic pleocytosis, without hypoglycorrhachia or elevated proteins. The presence of transient neurologic symptoms allowed to consider a possibly parainfectious mechanism, being the viral agent responsible for triggering the inflammatory process of the brain stem.

Conclusion

We describe the case of a girl with SARS-CoV-2 multisystemic inflammatory syndrome in children (MIS-C), with an unusual neurological complication, a parainfectious rhombencephalitis with a good clinical response to immunoglobulin treatment. The neurovirulence capacity of SARS-CoV-2 is only beginning to be recognized and long-term surveillance of these patients is required.

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Conflicts of Interest

The authors declare no conflicts of interest.

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