Short Communication

A sydenham chorea attack associated with COVID-19 infection

Merve Feyza Yüksel *, Miraç Yıldırım, Ömer Bektas, Süleymen Şahin, Serap Teber

Ankara University, Faculty of Medicine, Department of Pediatric Neurology, Turkey

1. Introduction

Severe acute respiratory syndrome coronavirus type 2 (SARS-CoV-2) caused by the coronavirus disease 2019 (COVID-19) has been spread all over the world since December 2019 and was accepted as a pandemic by the World Health Organization in March 2020. Until November 9, 2020, over 50,266,033 conﬁrmed cases and 1,254,567 deaths were reported ("Coronavirus disease 2019, 2020). Although COVID-19 primarily leads to symptoms in the respiratory and cardiovascular system, cases with neurological involvement have also been reported so far. COVID-19 associated neurological symptoms are more common in adult patients, while the number of cases with neurological involvement in children is limited. Neurological ﬁndings such as encephalopathy, seizures, headache, hypotonia, upward eye deviation, leg stiffening, cerebellar ataxia, muscle weakness, dysphagia, dystarthish have been detected in children, and no patients with chorea have been reported so far. In this article, we aim to present our case, which has been treated as Sydenham chorea, monitored without symptoms for the last six months and presented with chorea on the 3rd day of the COVID-19.

2. Case

A 14-year-old girl was admitted to our hospital due to the bending of the extremities and shoulder shrugging. Fifteen days before admission to the hospital, it was learned that her father was diagnosed with COVID-19. Five days after the contact, our patient developed shortness of breath and vomiting. She was tested positive for COVID-19 polymerase chain reaction (PCR) in the sample of nasopharyngeal swab, quarantined in the home without medical treatment. On the second day, the vomiting regressed. On the third day, the bending of her hands and feet, bilateral shoulder shrugging and discomfort started, and she admitted to the hospital on the 10th day of determining COVID-19 PCR positive. On the neurological examination, there were random, irregular and fast, involuntary choreiform movements on the bilateral (predominantly left) upper and lower extremities and bilateral milkmaid’s grip sign. Three years ago, she presented with chorea, at that time blood ceruloplasmin, thyroid function tests, anti-nuclear antibody (ANA), anti-double stranded DNA (anti-dsDNA), urine copper levels were detected normal, and cranial ultrasound revealed mitral and aortic valve insufficiency and she was diagnosed as Sydenham’s chorea. She treated with haloperidol for three months and then discontinued the drug due to improvement. The patient, who did not describe choreoathetosis for about 2 years, continued to have benzathine penicillin intramuscular schedule at intervals of 3 weeks. In the present attack, oral treatment of carbamazepine was started, and diagnostic survey were repeated. In laboratory examinations, glucose, electrolytes, liver-kidney-thyroid function tests, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), vitamin B12, and ceruloplasmin levels were found at normal ranges and iron deﬁciency anemia was detected. In serological tests, ANA and anti-dsDNA were negative.
antistreptolysin-O (ASO) was found to be 236 IU/mL (normal range: 0–200 IU/mL). The throat culture was negative for Group A β-hemolytic streptococci. Cranial MRI was normal. On the 7th day of admission, choreiform movements were improved.

3. Discussion

Although COVID-19 mostly affects the respiratory and cardiovascular system, neurological findings such as headache, dizziness, loss of smell and taste and neurological complications such as encephalopathy, acute cerebrovascular events, blurring of consciousness, and skeletal muscle damage have been reported in many cases since the beginning of pandemic (Mao et al., 2020; Helms et al., 2020).

COVID-19 is thought to cause neurological complications with two mechanisms: hypoxic brain damage and immune-mediated damage to the central nervous system. Systemic hypoxia, hypercarbia, and anaerobic metabolism caused by severe pneumonia lead to the accumulation of toxic metabolites, resulting in neuronal swelling and brain edema causing hypoxic brain damage (Tu et al., 2020). Immune-mediated damage depends on the activation of T lymphocyte, macrophages, and endothelial cells and the increase of inflammatory cytokines (Mehta et al., 2020; Tveito, 2020).

In the review of Ahmad et al. (Ahmad and Rathore, 2020), in which they published neurological findings and complications related to COVID-19 reported in the literature, dizziness, headache, acute cerebrovascular event, impaired consciousness, transverse myelitis, acute haemorrhagic necrotizing encephalopathy, encephalopathy, encephalitis and epilepsy were detected as central nervous system findings. Additionally, they reported peripheral nervous system findings and complications such as neuralgia, Guillain-Barré syndrome and skeletal muscle damage. These complications and symptoms were often seen in patients with severe clinical findings, in some cases, they appeared before respiratory and cardiac findings and may even be the sole finding.

On the other hand, Romero-Sánchez et al. (2020) showed that 57.4% of the patients developed at least one neurological complication in a study that analysed neurological manifestations during COVID-19. Nonspecific symptoms such as myalgia and headache were seen in the early stages of the disease and the meantime of occurrence of symptoms during this stage was 3.5 days. In the same study, neurological manifestations included nonspecific findings (myalgia, headache, dizziness, and syncope), symptoms associated with cranial nerves (loss of smell and taste), impaired consciousness (somnolence, stupor, coma, disorientation, and acute confusion syndrome), epilepsy (seizures and status epilepticus), peripheral nervous system manifestations (dysautonomia, acute inflammatory demyelinating polyneuropathy, muscle findings, hyperkalemia, rhabdomyolysis, and myopathy), cerebrovascular events (ischemic stroke and intracranial bleeding), hyperkinetic movement disorders, inflammatory findings (encephalitis and optic neuritis), and neuropsychiatric symptoms (anxiety depression, insomnia, and psychosis). Hyperkinetic movements observed in 6 patients on average 8.3 days after the onset of COVID-19 disease. Three patients with a history of neuropsychiatric disease had oromandibular dyskinesia, tremor in the upper extremities, and rigidity exacerbated by heavy pneumonia and neuroleptic use. The remaining three patients exhibited impaired consciousness that accompanied the myoclonic tremor holding the upper part of the body. There was no patient presenting with choreoathetosis or hypokinetic movement disorder. Likewise, two studies reported neurological findings and complications due to COVID-19 did not notify any movement disorders and choreoathetosis (Gkilos, 2020; Niazkar et al., 2020; Tveito, 2020).

In the literature, cases of neurological involvement associated with COVID-19 in children are rare but increase day by day. Abdel-Mannan et al. (2020) published neurological and radiological findings associated with COVID-19 in children and revealed brain stem involvement such as dystarthria and dysphagia, encephalopathy, headache, meningismus, and cerebellar ataxia in 4 previously healthy children. Peripheral nervous system involvement characterized by global proximal muscle weakness and decreased deep tendon reflexes were detected in all patients, two patients initially presented with neurological symptoms and the others exhibited neurological symptoms during follow-up. Moreover, upward eye deviation, leg stiffening (Chacon-Aguilar et al., 2020; Dugue et al., 2020), seizure (Bhatta et al., 2020), hypotonia (Nathan et al., 2020) and hypertonia (Vivanti et al., 2020) are other neurological findings reported in children previously.

Sydenham’s chorea is a condition that can show recurrence and its prevalence was reported as 42% in a study by Korn-Lubetzki et al. (2004), and 25% in another study (Carapetis and Currie, 1999). The recurrence of chorea is defined as the re-emergence of the new findings at least 2 months after the first attack and lasting longer than 24 h. It may occur months or years after the first chorea attack due to permanent subclinical damage to basal ganglia (Korn-Lubetzki et al., 2004). In a large cohort, Tumas et al. suggested that chorea recurrence in Sydenham chorea has been associated with irregular usage of prophylactic antibiotics (Tumas et al., 2007). On the other hand, some studies showed that patients with regular antibiotic prophylaxis also may suffer recurrences with unidentified immunological triggering factors (Korn-Lubetzki et al., 2004; Harrison et al., 2004). The current condition in the present patient may be exacerbated by Sydenham’s chorea or may be related to COVID-19 infection. It is noteworthy that symptoms appear on the 3rd day of COVID-19 infection, which may be associated with an immune mechanism triggered by a viral infection. The negative throat culture for Group A β-hemolytic streptococci and mild elevated ASO levels also can support this hypothesis. Moreover, she had no headache, seizure and impaired consciousness, or cranial MRI involvement, she had only pure choreiform movements. Although there have been previous reports of cases with COVID-19-related movement disorders in the literature, there is no case with chorea related COVID19. To the best of our knowledge, the present case is the first case with pure chorea reported during the course of COVID-19 in the literature.

4. Conclusion

Since December 2019, when SARS-CoV-2 first appeared, it has caused severe morbidity and mortality worldwide. Although primarily respiratory and cardiovascular system symptoms are seen, neurological involvement is also not uncommon and new cases of involvement and symptoms are reported day by day. In the pandemic process we experience, not only fever and respiratory symptoms, but also every atypical and unexpected symptom that occurs should be questioned in terms of COVID-19. Also, this case is contradictory to the patient samples in which COVID-19 infections create a cytokine storm and cause neurological involvement by severe inflammation. Even if the disease is mild, the virus may lead to a recurrence of diseases such as Sydenham chorea by reaching the central nervous system and trigger several local immunological events or reviving inflammation in the underlying stable sleep. This report expands the phenotypic spectrum of COVID-19 infection and suggests that COVID-19 infection can be associated with or trigger chorea. Further case reports are required to expand the phenotypic spectrum of COVID-19 infections.

References

Abdel-Mannan, O., Eyre, M., Löbel, U., et al., 2020. Neurologic and radiographic findings associated with COVID-19 infection in children. JAMA Neurol 77, 1–6. https://doi.org/10.1001/jamanetol.2020.2687.

Ahmad, J., Rathore, F.A., 2020. Neurological manifestations and complications of COVID-19: a literature review. J. Clin. Neurosci. 77, 8–12. https://doi.org/10.1016/j.jocn.2020.05.017.

Bhatta, S., Sayed, A., Ranabhat, B., et al., 2020. New-onset seizure as the only presentation in a child with COVID-19. Cureus 12, 8820. https://doi.org/10.7759/cureus.8820.

Carapetis, J.R., Currie, B.J., 1999. Rheumatic chorea in northern Australia: a clinical and epidemiological study. Arch. Dis. Child. 80, 353–358. https://doi.org/10.1136/adc.80.4.353.
Chacón-Aguilar, R., Osorio-Camara, J.M., Sanjurjo-Jimenez, I., et al., 2020. COVID-19: fever syndrome and neurological symptoms in a neonate. An. Pediatr. 92, 373–374. https://doi.org/10.1016/j.anpedi.2020.04.001.

Coronavirus Disease, 2020. Coronavirus disease 2019 (COVID-19) situation report. Available at: http://www.who.int. November 9.

Mehta, P., McAuley, D.F., Brown, M., et al., 2020. COVID-19: consider cytokine storm syndromes and immunosuppression. Lancet 395, 1033–1034. https://doi.org/10.1016/S0140-6736(20)30628-0.

Nathan, N., Prevost, B., Corvol, H., 2020. Atypical presentation of COVID-19 in young infants. Lancet 395, 1481. https://doi.org/10.1016/S0140-6736(20)30980-6.

Romero-Sánchez, C.M., Díaz-Maroto, I., Fernández-Díaz, E., et al., 2020. Neurologic manifestations in hospitalized patients with COVID-19: the ALBACOVID registry. Neurology 95, 1060–1070. https://doi.org/10.1212/WNL.0000000000009937.

Mehta, P., McAuley, D.F., Brown, M., et al., 2020. COVID-19: consider cytokine storm syndromes and immunosuppression. Lancet 395, 1033–1034. https://doi.org/10.1016/S0140-6736(20)30628-0.