A case of first pediatric pseudotumor cerebri syndrome secondary to superior sagittal sinus thrombosis associated with SARS-CoV-2

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INTRODUCTION

Pseudotumor cerebri syndrome (PTCS) is characterized by a constellation of symptoms due to elevated intracranial pressure with an unclear etiology and mandatory requirements of normal brain parenchyma and cerebrospinal fluid (CSF) constituents.[1] PTCS can be classified as primary and secondary depending on whether the etiologic agent is identified or not.[1] Intracranial venous thrombosis has been implicated as a cause for intracranial hypertension secondary to CSF outflow obstruction.[2]

Since the beginning of the pandemic, a diverse spectrum of neurological manifestations associated with SARS-CoV-2 infection, including headache, seizures, altered mental status, aseptic meningitis, and also intracranial hypertension has been identified.[3] Furthermore, SARS-CoV-2 infection deteriorate coagulation pathways, predisposing infected individuals to venous thromboembolis.[4] Underlying pathomechanisms include endothelial dysfunction with increased levels of von Willebrand factor, systemic inflammation with Toll-like receptor activation, and a procoagulatory state via tissue factor pathway activation.[4]

OBJECTİVES

We report a pediatric patient with secondary PTCS associated with sagittal sinus vein thrombosis after SARS-CoV-2 which, to our knowledge, has not been reported to date.

CASE PRESENTATION

A previously healthy 13-year-old boy presented to the emergency department with a history of headache, tinnutus and diplopia for the last four days. Neurological examination was normal except bilateral papilledema in fundoscopic evaluation. Detailed ophthalmologic examination revealed stage three papilledema, minimal dilation of the blind spot, bilateral 6 mm diameter of optic nerve sheath, increased peripapillary retinal nerve fiber layer thickness. Visual acuity was counting fingers from 4 meters. In the patient who had a history of SARS-CoV-2 infection 10 days ago, a SARS-CoV-2 polymerase chain reaction test from a nasopharyngeal swab was negative but SARS-CoV-2 was positive for IgM and IgG antibodies. Routine blood tests were normal. Brain magnetic resonance imaging (MRI) revealed partial empty sella, subarachnoid space enlargement of the bilateral perioptic nerve, and optic nerve tortuosity. MR venography revealed subacute dural venous sinus thrombosis in the short segment at the level of the superior sagittal sinus vertex (Fig.1). Echocardiogram, doppler ultrasonographies, genetic thrombophilia panel, hemostasis and rheumatological tests were normal. The opening pressure at the lumbar puncture was as high as 73 cm H2O. CSF biochemical parameters were normal. Extensive viral and bacterial serology was negative. Acetazolamide and low molecular weight heparin were started in the patient who was diagnosed with PTCS secondary to sagittal sinus vein thrombosis associated with SARS-CoV-2 infection. Due to metabolic acidosis and taste disturbance, acetazolamide was replaced with topiramate. On the third day of treatment, there was a significant improvement in headache and ocular complaints. Control ophthalmatological examination revealed normal visual acuity and peripapillary retinal nerve fiber layer thickness. Papilledema was regressed completely and bilateral optic nerve sheath diameters were 4 mm. The patient is still being followed up with topiramate and low molecular weight heparin.

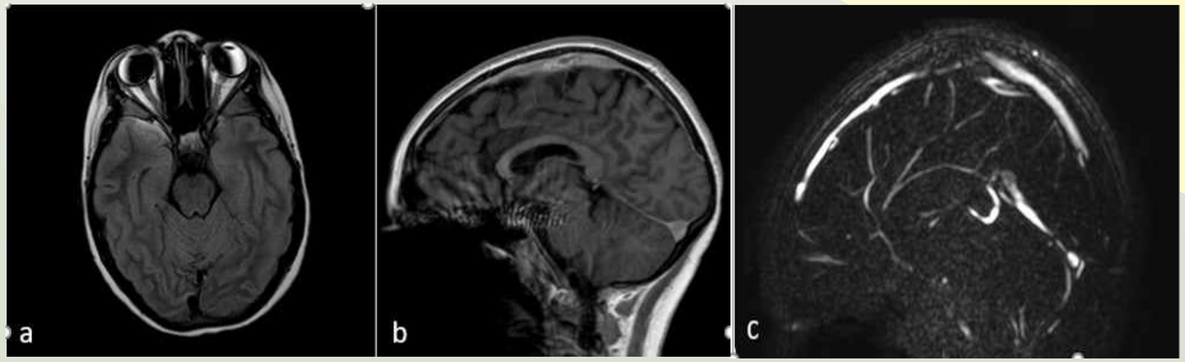

Fig. 1
a. Axial FLAİR image, shows protrusion of the optic nerve head and optic nerve tortousity.
b. Sagittal contrast-enhanced T1 image, focal hypointensity in superior sagittal sinüs due to partial thrombosis.
c. Sagittal TOF MR venography shows a lack of flow in the short part of the superior sagittal sinus

CONCLUSION

In patients with SARS-COV-2, presence of severe, persistent headache, isolated cranial nerve VI palsies, or papilledema should suggest secondary PTCS. Therefore, we recommend that in patients with confirmed SARS-COV-2 infection presenting unexplained neurological symptoms, a high degree of suspicion for CSVT should be always kept in mind.

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