Neurology Asia 2020; 25(4) : 607 – 610

COVID-19 infection presenting with acute thalamic infarction in a patient with Fahr's disease

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Abstract

Both Fahr's disease and SARS-CoV-2 infection can lead to thrombotic events. Fortunately, they are rare occurring together. There are no previous reports of SARS-CoV-2 infection in association with Fahr's disease. We report here a case of SARS-CoV-2 infection with Fahr's disease that presented with thalamic infarct. The patient improved with enoxaparin. Ischemic infarction may be seen in SARS-CoV-2 patients with associated Fahr's disease. Antithrombotic therapy may be helpful in preventing further deterioration.

Keywords: COVID 19, Fahr's Disease, SARS-CoV-2 infection, Thalamic infarct

INTRODUCTION

Severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infections, which often occur with respiratory signs, can rarely affect the nervous system. Different clinical manifestations, such as dizziness, ataxia, epileptic seizures, ischemic stroke, sinus vein thrombosis, cerebral hemorrhages, encephalitis, meningitis and radiculitis may be seen.[1,2] It has been reported that a predisposition to arterial and venous thrombosis can also occur with ischemic and hypoxic encephalopathy.[3-5]

Fahr's disease (familial idiopathic basal ganglia calcification) is characterized by calcifications localized to the bilateral basal ganglia, dentate nucleus, centrum semiovale and cerebellum. With a predisposition to ischemic phenomena, it can present with extrapyramidal symptoms, cerebellar dysfunction, speech impairment, dementia, and neuropsychiatric symptoms. There is also predisposition to ischemic stroke.[6,7]

There are no previous reports of SARS-CoV-2 infection with Fahr's disease. We present here a case of SARS-CoV-2 infection with Fahr's disease with thalamic infarct confimed by imaging studies.

CASE REPORT

A 48-year-old male was admitted to our emergency service with complaints of nausea, vomiting and headaches. The patient had no significant abnormality on physical examination, and no abnormality was detected in the thoracic computed tomography (CT) (Figure 1a). Non-contrast brain CT revealed only symmetrical calcifications in bilateral basal ganglia and dentate nucleus, which are indicative of Fahr's disease, and no acute hemorrhagic or ischemic pathological manifestations were observed (Figure 1b).

Laboratory examinations revealed elevation in only blood urea nitrogen (BUN) and creatinine levels [BUN:143 mg/dl (NR 17-43), Creatinine:2,07 mg/dl (NR 0,67-1,17)]. After symptomatic treatment, the patient recovered and was discharged.

Two days after the first admission, persistent nausea and vomiting increased and the patient also developed somnolence, respiratory distress and dyspnea. The SpO_2 of the patient was 80%–82% without oxygen support, and thoracic CT examination revealed diffuse interlobular septal thickenings and nodular ground-glass appearance (Figure 2a). While D-Dimer [6340 μg/L (NR 0-654)], ferritine [1828 μg/L (NR 23,9-336)], CRP [38 mg/L (NR 0-5)] increased at blood test, haemoglobin [9,6 g/dl (NR 12,5- 16,3)], haemotocrit [%27,5 (NR % 36,7-47,1)] and white blood cells [3,2x10^3/μl (NR 3,6-10,2)] values decreased.

With these clinical, laboratorial and radiological findings, the patient was hospitalized in the intensive care unit with the preliminary diagnosis

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Date of Submission: 1 July 2020; Date of Acceptance: 10 August 2020

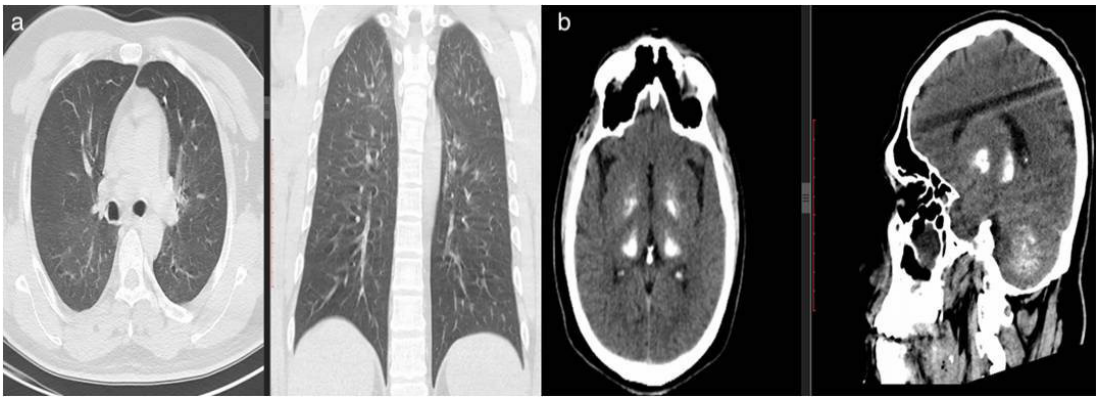

Figure1. **a** Thoracic computed tomography at first admission was normal, **b** Non-contrast brain CT shows bilateral basal ganglia and dentate nucleus symmetrical calcifications

of SARS-CoV-2 infection. Nasopharyngeal swab sampling result was positive of coronavirus disease 2019 (COVID-19).

Neurological examination showed that the patient was drowsy, with impaired cooperation and orientation. The Glasgow coma scale of the patient was 13. On subsequent follow-up, it was observed that the place and time orientation was impaired and patient developed tremor. The nausea and vomiting subsequently improved with medical treatment. Oxygen support was given through the nasal cannula. Hydroxychloroquine 2 × 200 mg (5 days, oral administration), levofloxacin 1 × 500 mg (intravenous administration), and enoxaparin 2 × IU/0.4 mL (subcutaneous administration) were started as part of the medical treatment.

Bilateral thalamic hyperintensities consistent with infarct was seen in the cerebral magnetic resonance imaging (MRI) and diffusion MRI (Figure 2b, c). The cerebrospinal fluid examination was not supportive of encephalitis. (CSF results: Glucose 55 mg/dl (NR:45-80), Na: 135 mEq/L (NR: 135-155), protein 38 mg/dl (NR 15-45) and no leukocyte)

After 1 week of intensive care treatment, oxygen saturation was normalized. Neurological examination showed no abnormality. Radiological pathological findings detected by thoracic imaging at admission significantly improved (Figure 3a). Repeat cerebral MRI examination on 10th day showed partial regression in the thalamic region ischemic infarction (Figure 3b).

The patient was discharged after the quarantine period of 15 days was completed.

DISCUSSION

SARS-CoV-2 infections may manifest with polyneuropathy, encephalopathy, ischemic stroke, paralysis, paraesthesia, and epileptic seizures.[8]

It has been suggested that anaerobic metabolic abnormalities due to respiratory distress in patients with SARS-CoV-2 infections may result in neurological damage.[9,10] Thrombotic phenomena with hypercoagulability is commonly observed in COVID-19 infected patients.[11]

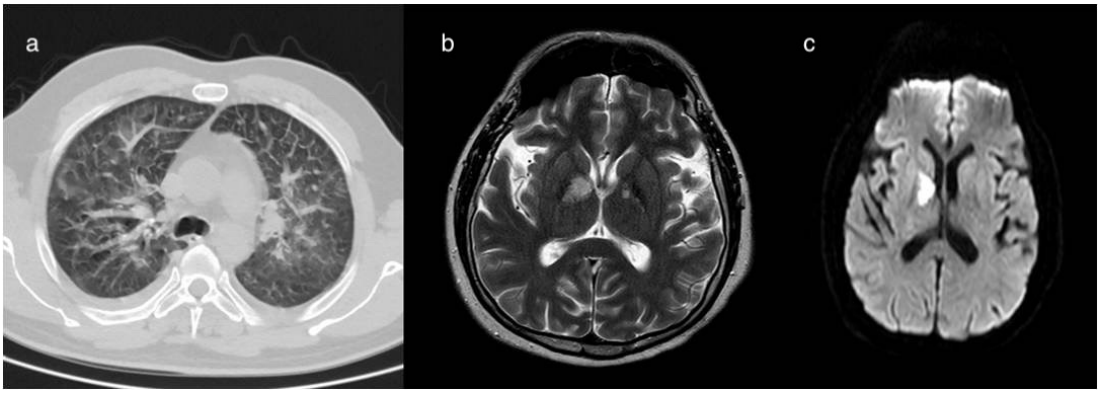

Figure 2. **a** Thoracic computed tomography at second admission, **b**, **c** Cerebral magnetic resonance imaging (MRI) and diffusion MRI

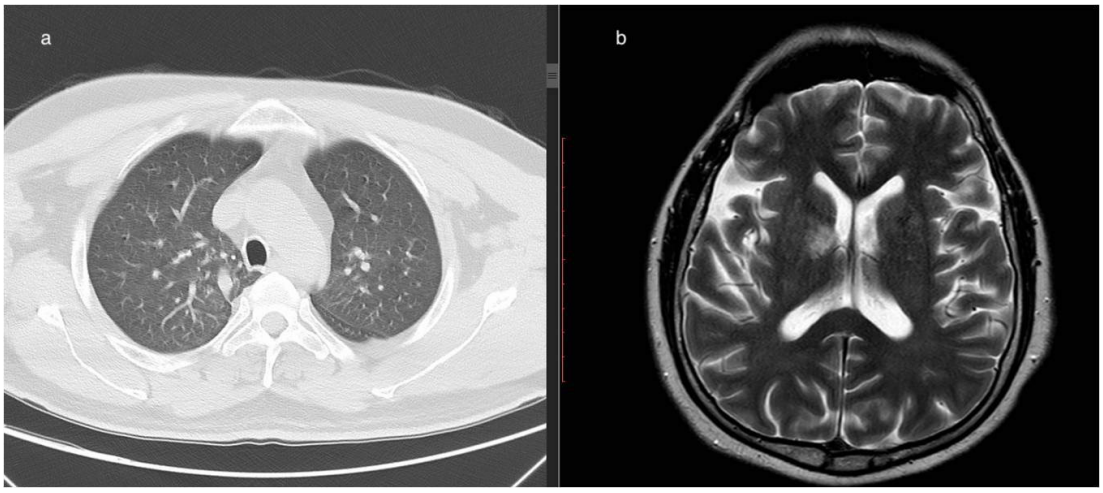

Figure 3. **a** Thoracic computed tomography at discharge, **b** Control cerebral MRI examination showed partial regression in the thalamic region ischemic infarction

In the study by Lu *et al*., ischemic neurological incidents were observed in 27% of 304 COVID-19 patients.[12] In another study on 184 COVID-19 patients, cerebrovascular events were responsible for 10.8% of the 92 patients who died.[13]

In another study, approximately 20% of 113 SARS-CoV-2-infected patients had hypoxic, ischemic encephalopathy.[3] In a Chinese study, 214 patients were evaluated and 36% of patients had acute ischemic and hemorrhagic cerebrovascular pathology.[4] Agarwal *et al*. reported that SARS-CoV-2 infection is associated with increased risk of intracerebral haemorrhage.[5] Therefore, we avoided aggressive thrombolytic therapy. We used low molecular weight heparin for the treatment.

Fahr's disease was described by Theodore Fahr in 1930 as "idiopathic calcification of cerebral vessels." It is often seen in individuals between 40 and 60 years of age, and its prevalence is <1/1,000,000. There is a predisposition to thrombotic and ischemic phenomena, which may present with neurologic and neuropsychiatric symptoms.[6,7] Fahr's disease is marked by the accumulation of various minerals, glycoproteins, or mucopolysaccharides underlined by regional ischemia or inflammation. It is triggered by antiepileptic drugs and radiotherapy affecting the calcium metabolism.[14] Due to these changes in the vascular structure, cerebral blood flow can be decreased and thrombotic phenomena may develop. Therefore, ischemic hypoxic and hemorrhagic encephalopathy can be seen in Fahr's disease.

In conclusion, SARS-CoV-2 infection and Fahr's disease can both lead to neurological involvement in the form of thrombotic, ischemic phenomena. We report here a patient with COVID-19 who also had underling Fahr's disease. The patient developed thalamic infarct.

DISCLOSURE

Conflict of interest: None

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