Journal of the Neurological Sciences 400 (2019) 21–22

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Letter to the Editor

A case of cerebral infarction in a patient with TAFRO syndrome

ARTICLE INFO

Keywords:
Cerebral infarction
TAFRO syndrome
Disseminated intravascular coagulation
Anti-inflammatory therapies

Dear Editor,

A systemic inflammatory disorder, TAFRO syndrome is characterized by thrombocytopenia, anasarca, fever, reticulin fibrosis and organomegaly. It was first described in 2010 [1], and similar cases have been reported since then [2]. The pathophysiology of TAFRO syndrome is not fully understood, and various clinical manifestations have been reported. To our knowledge, there is no report of ischemic complications in patients with TAFRO syndrome. Here, we report a case of a patient who developed cerebral infarction during the clinical course of TAFRO syndrome.

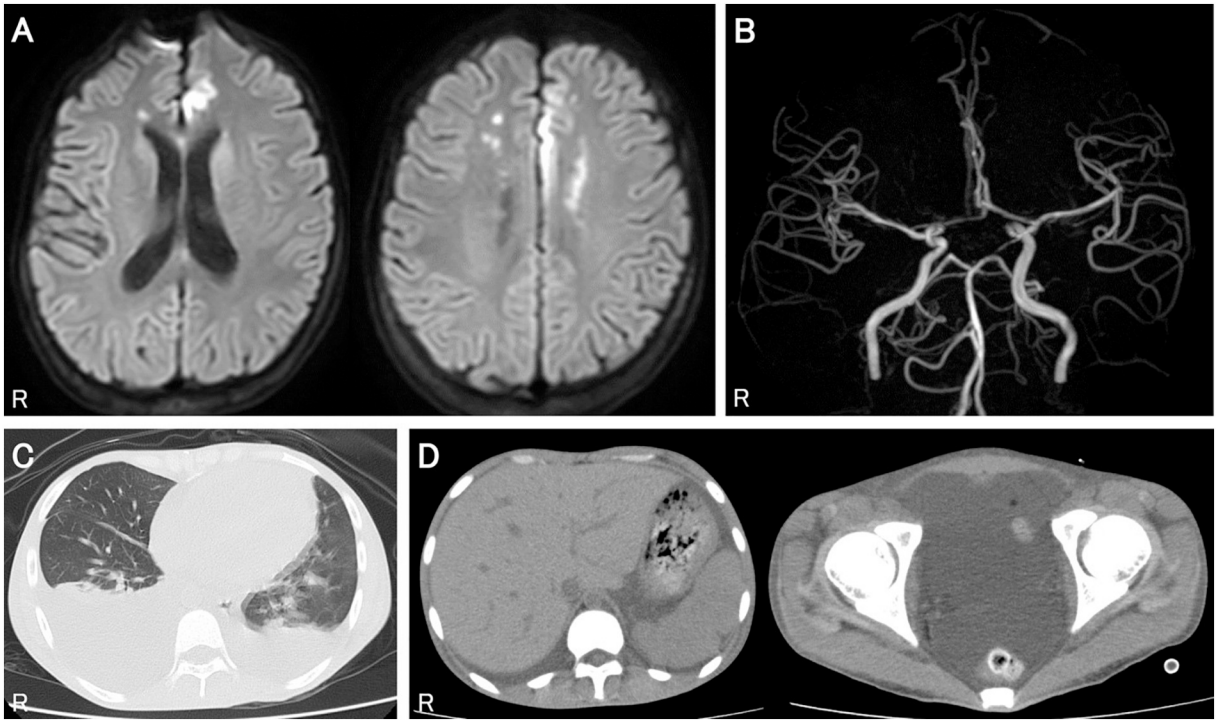

Fig. 1. Radiological findings on admission. (A) Brain magnetic resonance imaging showing high-signal-intensity areas in the bilateral anterior cerebral artery region on diffusion-weighted images. (B) Brain magnetic resonance angiography showing no abnormal findings. (C) Chest computed tomography (CT) showing bilateral pleural effusion. (D) Abdominal CT showing ascites and mild splenomegaly.

https://doi.org/10.1016/j.jns.2019.03.006
Received 15 December 2018; Received in revised form 3 March 2019; Accepted 11 March 2019
Available online 12 March 2019
0022-510X/

1. Case presentation

A 17-year-old man was admitted to our hospital with mild disturbance of consciousness and weakness of limbs. He had a medical history of repaired Tetralogy of Fallot and Goldenhar syndrome. On admission, he had a Glasgow Coma Scale of 15, but was found to be slightly drowsy. Physical examination revealed anasarca and mild muscle weakness in both lower limbs. No other neurological abnormality was observed. Blood tests revealed thrombocytopenia (39,000/μL); normal level of immunoglobulin G (1,292 mg/dL); prolonged prothrombin time (15.3 s); and elevated levels of creatinine (1.09 mg/dL), C-reactive protein (12.08 mg/dL), alkaline phosphatase (721 U/L), D-dimer (44.9 μg/mL), interleukin-6 (11.3 pg/mL) and vascular endothelial growth factor (206.0 pg/mL). He was diagnosed with disseminated intravascular coagulation (DIC) based on the International Society of Thrombosis and Hemostasis criteria [3]. Brain magnetic resonance imaging revealed high-signal-intensity areas in the bilateral anterior cerebral artery region on diffusion-weighted images (Fig. 1A). Brain magnetic resonance angiography showed no stenotic lesion or encasement in the bilateral anterior cerebral artery (Fig. 1B). Computed tomography revealed bilateral pleural effusion, ascites, and mild splenomegaly (Fig. 1C and D). Bone marrow biopsy revealed slight reticulin fibrosis. To investigate the embolic source, carotid artery ultrasonography, transthoracic echocardiography, lower limb vein ultrasonography, and Holter electrocardiography were performed, but yielded no abnormal finding. Renal dysfunction had progressed after admission, and the serum creatinine level increased to 1.50 mg/dL on day 5. We diagnosed the patient with TAFRO syndrome based on the clinical findings of anasarca, thrombocytopenia, systemic inflammation, organomegaly, renal dysfunction, absence of hypergammaglobulinemia, and high level of alkaline phosphatase [4,5]. After initiation of anti-inflammatory therapies including oral prednisolone (1 mg/kg/day, followed by tapering), cerebral infarction did not recur. He was discharged on day 51 without neurological deficit.

2. Discussion

This is the first case report of a patient with TAFRO syndrome who developed cerebral infarction. The etiology of cerebral infarction is unknown. We speculate that the thrombus resulting from DIC might be the embolic source. Another potential pathogenic mechanism is that prolonged intravascular hypovolemia due to increased systemic vascular permeability might cause cerebral ischemia.

The symptoms of TAFRO syndrome are the results of a cytokine storm, the overproduction of inflammatory cytokines by immune system [4,6]. The induced cytokine storm results in increased systemic vascular permeability, leading to multiple organ dysfunction. In addition, some cases of TAFRO syndrome are complicated by DIC [7]. The pathophysiological mechanisms of DIC in TAFRO syndrome is uncertain, but it may also occur due to a cytokine storm. Anti-inflammatory drugs, such as glucocorticoids, cyclosporin A, tocilizumab, and rituximab, are reported to be effective for treatment of TAFRO syndrome [4]. These treatments might help prevent recurrence of cerebral infarction associated with TAFRO syndrome.

In conclusion, clinicians should be aware that ischemic complications might manifest in TAFRO syndrome.

Conflicts of interest

None.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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