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Steroid Responsive Encephalopathy Associated with Autoantibodies to Thyroperoxidase (STREAT), Presenting with Acute Stroke in a Young Female Patient

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Case Summary

A young lady in her mid-30s was presented with acute ischemic stroke, associated with encephalopathy, in the form of confusion and agitation. Clinical examination demonstrated a profound tachycardia and thyroid swelling. Subsequent investigations revealed a multinodular goiter, with elevated thyroperoxidase antibodies (TPO Ab), on serology. Treatment with high-dose intravenous corticosteroids leads to a prompt remission of the disease and related encephalopathy, and a dramatic clinical improvement in the patient.

Background

This is a rare presentation of steroid responsive encephalopathy associated with autoantibodies to thyroperoxidase (STREAT), in a young female patient. Owing to the rarity of this condition, diagnosis can be missed, leading to delay in initiating treatment and an eventual catastrophic outcome. However, if diagnosed promptly, steroid therapy usually leads to a dramatic clinical improvement.

Case Presentation

A lady in her mid-30s presented with an acute onset of right-sided weakness, right upper motor neuron facial weakness, pseudobulbar palsy, and expressive aphasia. This was associated with confusion, agitation, a decreased attention span, mood disturbance, being emotionally labile, and insomnia. Her husband reported that she had recent weight loss, and intolerance to hot weather. In addition to her neurological status, on examination she was underweight, and tachycardiac with observable thyroid swelling. Her temperature and blood pressure were within normal limits.

Methods

On admission, MRI of the brain with diffusion weighted images (DWI) sequence showed an area of restricted diffusion of the left basal ganglia, internal capsule, operculum, with a corresponding hypodensity in the apparent diffusion coefficient (ADC) map; consistent with a recent acute ischemic infarction of the left middle cerebral artery distribution (Figure 1).

Routine biochemical and hematological investigations were normal apart from a moderate microcytic hypochromic anemia (Hb = 9.5, MCV = 76, MCH = 22, MCHC= 30). Remarkably, on serology, markers for autoimmune vasculitis and hypercoagulable states were negative.

On cardiac assessment, the ECG demonstrated a sinus tachycardia with normal transthoracic and transesophageal echocardiography (TTE and TEE, respectivley).

On thyroid assessment, a thyroid ultrasound showed a multinodular goitre. A thyroid profile showed low thyroid stimulating hormone (TSH = 0.01 uIU/ml), with high free-T3 (12.5 pg/ml) and T4 (5 ng/dl). TPO Ab were markedly elevated (1615 IU/ml, normal reference < 50 IU/ml).

Treatment and Outcome

Our treatment plan consisted of high dose (1 g) intravenous methyl-prednisolone for 5 days, followed up with bridging to 60 mg oral prednisolone/day.

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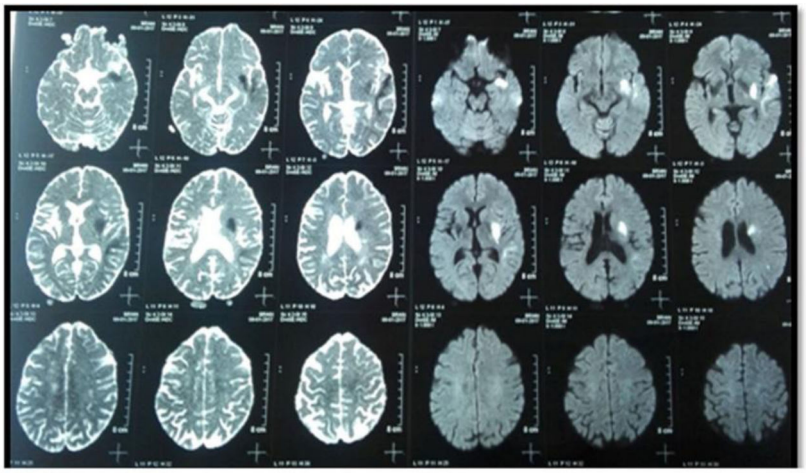

Figure 1. Brain MRI with DWI sequence showed an area of restricted diffusion of the left basal ganglia, internal capsule, operculum, with a corresponding hypodensity in the ADC map.

Following up from the treatment, the patient showed a dramatic improvement in the neuropsychiatric manifestations and tachycardia, with a remarkable regression of the National Institute of Health Stroke Scale/Score from 18 to 6. This improvement showed the tremendous effect of steroids in treating this patient. The patient was followed up in clinic four months later, to assess recovery. Positively, she was adherent to steroid therapy throughout this time and had an eventual modified Rankin score of 1, with an excellent functional outcome.

Discussion

In this case of a young female lady presenting with acute stroke with a deep territorial infarct, we attempted to exclude other possible underlying etiologies.

The first differential diagnosis suspected was cardioembolism due to a territorial distribution of the cerebral infarcts and the fact that the patient had a demonstrable tachycardia on admission. However, during the period of hospital stay and with Holter monitoring, the rhythm demonstrated was a persistent sinus tachycardia, without any other underlying arrhythmias. Echocardiogram (TTE and TEE) was normal, with normal visualization of the valvular, septal, and myocardial morphology. Cardiac contractility was also normal with average-sized cardiac chambers, for age and sex.

Second, antiphospholipid syndrome was suspected owing to a history of repeated abortions, on three occasions. Further clinical history clarified that these abortions were nonconsecutive, without any other obstetric history of premature birth, severe preeclapmia, eclampsia, or placental insufficiency. Laboratory markers for antiphospholipid syndrome (lupus anticoagulant, anticardiolipin, anti-β2 glycoprotien) were all shown to be negative. Thus, this did not fulfill Sydney's criteria for antiphospholipid syndrome diagnosis [1].

We also considered other immune-mediated vasculitides such as Systemic Lupus Erythematous (SLE), Wegner's granulomatosis. The patient also did not fulfill the diagnostic criteria for either of these conditions. The irrespective diagnostic laboratory markers, such as ANA, Anti-dsDNA and ANCA, all returned negative.

Thrombophilia was also excluded by laboratory markers for hypercoagulable states, such as protein C, S, antithrombin III activity, and a thrombophillia gene profile, all returning negative.

In the presence of demonstrable thyroid swelling, associated with encephalopathic manifestations, this promoted further investigations for the thyroid gland, which were obviously positive for hyperthyroidism. Hyperthyroidism explained the recent weight loss, heat intolerance and tachycardia. In addition, together with the observation of high titer of TPO Ab, the diagnosis was consistent with Hashimoto thyroidits.

The current case represents a less common presentation of a rare neuro-endocrinal disorder. STREAT has a broad range of clinical manifestations. The mainstay of diagnosis is the presentation of patients with encephalopathy, CNS dysfunction, high titer of thyroid antibodies, and a good therapeutic response to immunosuppressants, particularly steroids [2].

The exact pathogenic mechanism underlying the disease is unknown, but an immune-mediated response against common brain–thyroid antigens is currently proposed. Some studies have described the underlying pathogenic process as vasculitic [3,4], while others have denoted a nonvasculitic component, with meningeal and parenchymal brain involvement [5].

Generally, the neuro-psychiatric manifestations have been described in two distinct forms: a vasculitic type, characterized by multiple relapsing-remitting stroke-like episodes, and mild cognitive impairment (as in our patient) or a diffuse progressive type, of which the cardinal features are dementia and psychiatric symptoms. Alterations in the consciousness level (stupor or coma), tremors, seizures, or myoclonus can occur with either forms [6].

This disorder is a disease of middle aged females, with a female:male preponderance, and ratio of 5:1, with a mean age of onset at 45–55 years of age. However, both pediatric and late-onset cases have been reported in the literature [7].

In our case of a young female patient, in her mid-30s with a presentation of acute stroke and a profound encephalopathy from onset; an exclusion of any known causes of stroke in young patients with appropriate investigations; in addition to a very high titer of TPO Ab (1615 IU/ml, normal reference < 50 IU/ml), and subsequent dramatic response to steroids, objectively confirms the diagnosis of STREAT induced acute stroke.

Finally, high clinical index, the prompt recognition, diagnosis and appropriate treatment of STREAT can dramatically improve clinical and functional outcomes in this subset of patients.

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