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Diffuse Systemic Sclerosis with Brainstem Encephalitis: A Case Report

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Abstract

Brainstem encephalitis (BE) is a rare disease that affects 1 in 10,000 individuals each year. However, quite little is known about the aetiologies causing brainstem encephalitis. Scleroderma is a rare connective tissue disorder with pathophysiology still under research. We hereby report a case of systemic sclerosis that primarily presented with brainstem encephalitis, a relatively uncommon presentation, and was retrogradely evaluated, assessed, and diagnosed as diffuse systemic sclerosis (Ds-SSc). Systemic sclerosis is associated with multisystem involvement (multiple internal organs simultaneously or sequentially) and is associated with increased morbidity and mortality, with uncommon involvement of the brainstem. Hence, in any case of brainstem encephalitis in a young patient with no apparent infectious cause, an elaborate examination of all systems and autoimmune profiles must be performed to enable rapid initiation of treatment and, therefore, improve prognosis.

Keywords: Rhombencephalitis; Brainstem encephalitis; Scleroderma; Systemic sclerosis.

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Introduction

Brainstem encephalitis (BE) is a rare condition with a prevalence of around 1 in 10,000[1]. Most cases of BE are idiopathic. Among the patients with identifiable etiologies, infections remain the most common cause[2]. Still under active research, the spectrum of causes and outcomes of BE has not been systematically studied[3]. With limited case reports and a series of resources, Bickerstaff's brainstem encephalitis, Listeria rhombencephalitis, and Ma2-associated paraneoplastic encephalitis are among the common entities described in BE. Systemic sclerosis is associated with multisystem involvement (multiple internal organs simultaneously or sequentially) and is associated with increased morbidity and mortality, with uncommon involvement of the brainstem.

Hereby, we report a case of systemic sclerosis that primarily presented to us with brainstem encephalitis, a rather uncommon presentation that was retrogradely evaluated and assessed, and the diagnosed case of diffuse systemic sclerosis (DS-SSc).

Case Report

A 33-year-old male presented to us with altered behavior and irrelevant talk for the past 4 days. The patient was asymptomatic 4 days back when he woke up one morning and started behaving irrationally with increased agitation and aggressiveness, with episodes of running away from home due to restlessness and abuse of attendants when trying to control him. During the episode, the patient had no difficulty remembering his family, home, address, date, and time. No history of fever or self-harm. A local doctor prescribed him selective serotonin reuptake inhibitors (SSRIs), Quetiapine, and Benzodiazepines, but with partial relief. He had recurrent episodes of generalized tonic-clonic seizures. He was then brought to our emergency department.

The attendants report a history of darkening of the skin, symmetrically diffuse over the face and body, gradually worsening over the past 1 year. The patient also complained of an inability to open, resulting in discomfort during thorough chewing. He felt more tightness and dryness in his eyes and face over the past 6 months. Over the past 6 months, he also complained of symmetric polyarthritis, breathlessness on exertion for the past 4 months, and loose stools for the past 3 months. He also had difficulty walking with the imbalance and a tendency to fall on either side, sometimes for the past 3 months. He has been diagnosed with hypertension for the past 3 months.

Upon examination, he had a salt-and-pepper type of hyperpigmentation on his face, trunk, and all limbs [Figure 4]. His skin appeared stretched and shiny, and his

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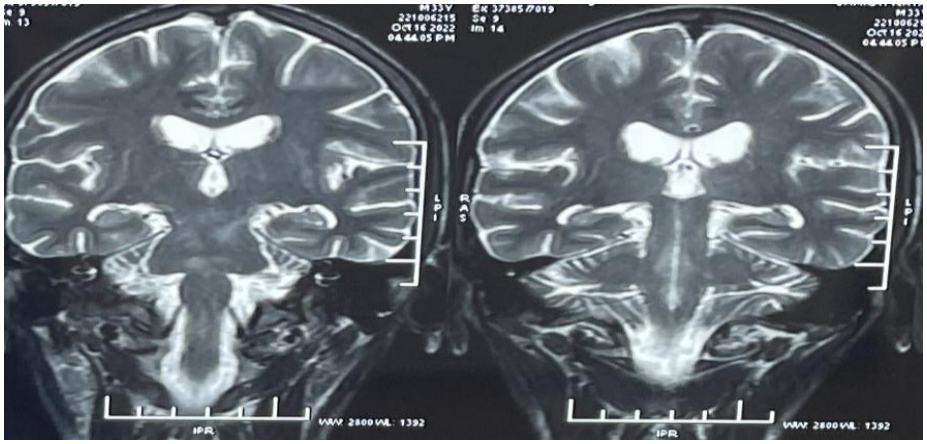

Figure 1. T2-Weighted MRI Brain (coronal section) showing brainstem midline hyperintensity

oral fissure was narrow with sharp angles. Modified rodnon skin score (MRSS) was 32/51. He was then admitted to us for further evaluation. His serum complete blood count, peripheral blood smear,serum electrolytes (Sodium, Potassium, Calcium), liver function tests, and urine routine microscopy were sent, which were non-significant. His serum anti-myelin oligodendrocyte glycoprotein (anti-MOG) and anti-aquaporin-4 were sent and reported negative. Having both integumentary and neurological system involvement, his anti-nuclear antibody (ANA) titer was 4+ with a finely speckled appearance. So, the patient's serum ANA profile was sent. It suggested that anti-RO and anti-ribonucleoprotein (anti-RNP) antibodies were significantly positive. His fundus was normal. His brain magnetic resonance imaging (MRI) was done, which suggested brainstem encephalitis [Figure 1-3]. His serum thyroid-stimulating hormone (S.TSH) was 10.20, T3 (Free) 42.56, T4 (Free) 4.83, C-reactive protein (CRP) levels- 6.98, erythrocyte sedimentation rate (ESR)- 125, blood urea- 96, serum creatinine (S. Creat)– 2.20. His 2D echocardiography (ECHO) showed global hypokinesia with an ejection fraction (EF) of 48%. His high-resolution computed tomography (HRCT) chest showed linear and reticular interstitial opacities bilaterally.

Hence, the patient was started on i/v high-dose steroids (i.e., 1gm Methylprednisolone) for 5 days. The patient showed significant improvement in sensorium with almost normal behavior by DAY-3. The patient was discharged on a maintenance steroid dose (i.e., 1mg/kg/day tapered by 10mg every 1 week over 8 weeks) with calcium supplementation.

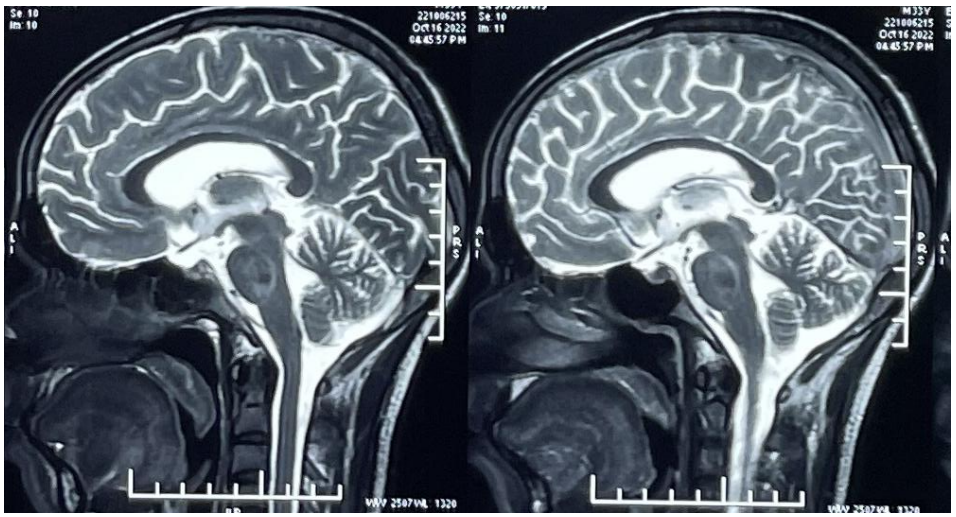

Figure 2. T2-Weighted MRI Brain (sagittal section) showing brainstem midline hyperintensity.

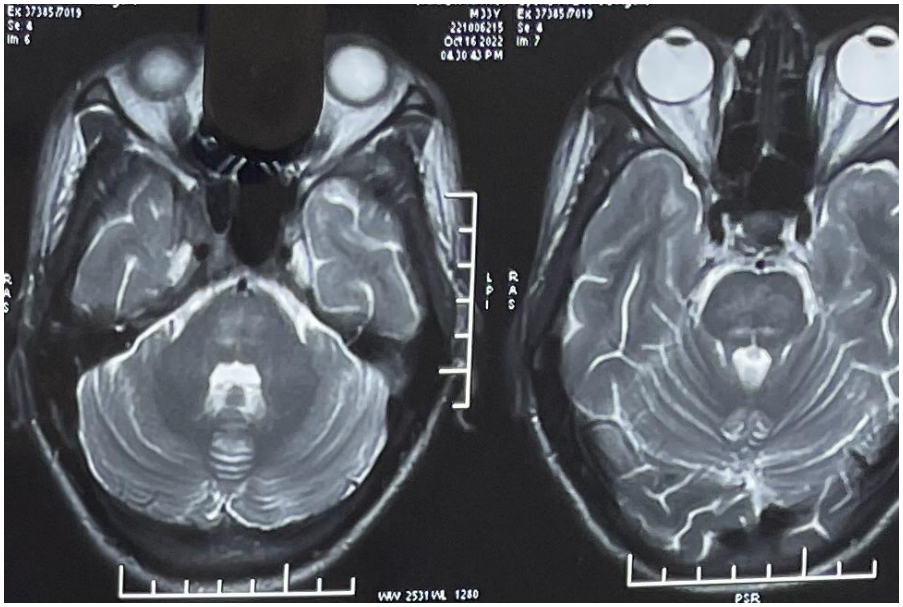

Figure 3. T2-Weighted MRI Brain (axial section) showing brainstem midline hyperintensity.

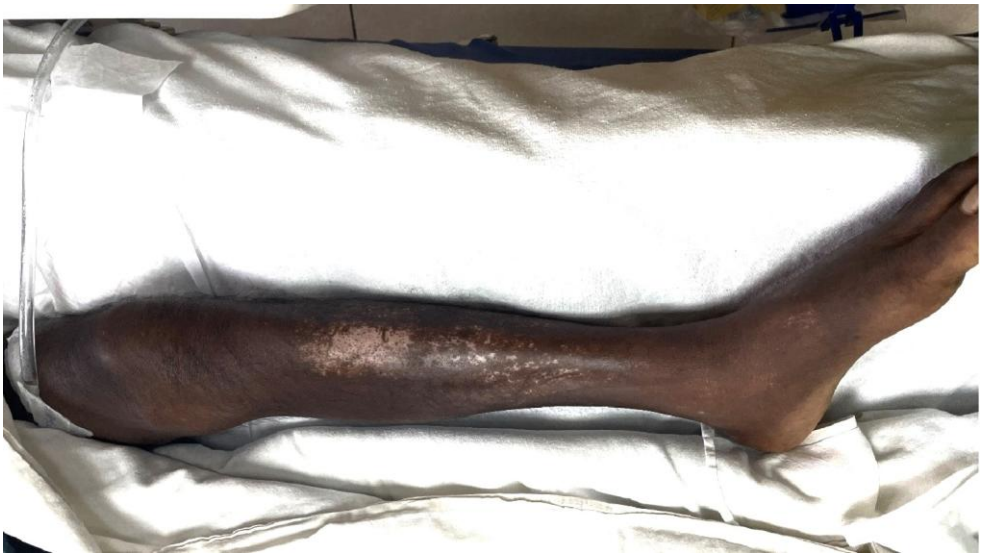

Figure 4. Diffuse salt & pepper hyperpigmentation of skin.

Discussion

Under many non-infectious aetiologies described, autoimmune brainstem encephalitis has a rare presentation. It can be divided into two broad categories: primary central nervous system (CNS) inflammatory disease, or secondary to systemic diseases in which the CNS is one of the affected systems[7]. Diffuse cutaneous systemic sclerosis (DcSSc) is characterized by thickening of the skin on the proximal extremities, face, and trunk, and is associated with several systemic manifestations. It shows autoantibodies in the serum, specifically antinuclear antibodies (ANA), which are present in more than 90% of cases. Out of the more specific autoantibodies (anti-centromere, anti-SCL70, and anti-RNA polymerase III), at least one of them is positive in up to 70% of cases. The most common organs affected are the skin, gastrointestinal tract, lungs, kidneys, skeletal muscle, and pericardium[4,5,6].

The reported case was diagnosed clinically and radiologically as brainstem encephalitis. Differential diagnoses of psychosis, metabolic encephalopathy, HSV or other infectious encephalitis, and white matter disease were considered. Infectious aetiologies were ruled out based on clinical and laboratory parameters, and inflammatory pathology for the same was suspected

because he had polyarthralgia, diffuse hyperpigmentation, dyspnoea, and CNS manifestations. Based on his dermal and respiratory symptomatology, specific laboratory tests were sent to find the cause of inflammation and autoimmune multi-systemic disease, with an ANA profile suggesting Diffuse systemic sclerosis. He was counselled, and treatment with solely corticosteroids was significantly successful.works and connects in people with ADHD, focusing on specific frequency bands. The study's results were quite impressive, with a 99.8% accuracy in the γ band across the entire brain. Notably, the right hemisphere, particularly in the θ band, showed significant differences between ADHD and HC, resulting in 98.5% accuracy.

These findings suggest that the right hemisphere plays a crucial role in ADHD symptoms.

Future research directions could involve scaling this methodology to larger datasets and incorporating diverse cognitive tasks, potentially uncovering deeper insights into brain interactions. The success of this approach, combining streamlined architecture with color-coded connectivity images, holds promise for real-world applications and cost-effective diagnostic tools.

Conclusion

Hence, brainstem encephalitis may be a complication of systemic sclerosis. In cases of brainstem encephalitis in young patients with no apparent infectious cause, an extensive evaluation of all systems and autoimmune profiles must be performed to enable rapid initiation of treatment and, therefore, improve prognosis.

Acknowledgments

None.

Competing Interests

The authors declare no conflict of interest.

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None.

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