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Case Report

A novel case of rickettsial encephalitis presenting as isolated global aphasia

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ABSTRACT

This case report details a novel presentation of rickettsial encephalitis in a 25-year-old male patient who arrived with a 4-day history of fever and altered sensorium, marked by no verbal output and a loss of social inhibitions. Clinical examination revealed a 2x2 cm eschar on the right leg, moderate splenomegaly, thrombocytopenia, and leucopenia. An MRI of the brain showed focal T2/FLAIR hyperintensity involving the splenium of the corpus callosum, suggestive of cytotoxic lesions of the corpus callosum (CLOCCs). Despite the lack of structural abnormalities in primary language areas, the patient exhibited global aphasia. A positive Weil-Felix test indicated rickettsial infection. The patient was treated with ceftriaxone and doxycycline, leading to gradual neurological improvement and complete recovery within 15 days. This case highlights the importance of high clinical suspicion for rickettsial encephalitis, timely diagnosis, and appropriate antimicrobial therapy to achieve favorable outcomes.

Keywords: Rickettsial fever, Encephalitis, CLOCCS

INTRODUCTION

Rickettsial infections are the vector borne diseases re-emerging worldwide. Although commonly, a triad of fever, cutaneous rash and an eschar (inoculation site) is typically seen in rickettsiosis, these diseases can have an array of manifestations involving various organ systems including CNS.[1] The most common CNS presentations of rickettsial infections are headache, behavioral abnormalities, focal neurological deficits, seizures, meningitis and meningoencephalitis and coma.[1] Lately, cases of infectious cerebral vasculitis, acute ischemic stroke, unilateral facial nerve palsy, pure alexia syndrome, etc have been reported with rickettsial fever.[2-4] However, low index of suspicion by the physicians and lack of easy availability of diagnostic tests leave rickettsial infections undiagnosed.[5,6] This increases morbidity and mortality of these infections even though timely use of appropriate antimicrobials can have good outcome. Our case report illustrates an interesting case of 25-year-old man who was presented with global aphasia, with peculiar findings on the MRI brain, who was diagnosed to have rickettsial encephalitis.

CASE REPORT

A 25 years old male security guard was brought to Goa medical college casualty with complains of fever for 4 days and altered sensorium in form of no verbal output, no relevant responses to verbal commands, loss of social inhibitions since, two days. He was found in his own room in this state. He had no history of any chronic illnesses in the past. His co-workers denied any history of nausea, vomiting, visual disturbances, seizures in the patient. He used to occasionally consume alcohol but there was no history of any recent binges of alcohol. On examination, patient's vitals were normal. A 2×2 cm lesion was noted on right leg suggestive of an eschar. On PA examination, there was moderate splenomegaly. Patient was conscious, spontaneously opening eyes, moving all 4 limbs however

comprehension was absent. Patient was aphasic. Tone was increased in all limbs. Neck stiffness was absent. Deep tendon reflexes were brisk in all joints. Bilateral planters were flexors. Patient's CBC revealed thrombocytopenia with platelet counts of 18000 and leucopenia with TLC 1100. Total kidney and liver function tests, serum electrolytes, urine routine and blood culture did not show any significant abnormality. CT brain was normal. Ultrasound of the abdomen revealed minimal free fluid in the abdomen suggestive of inflammation.

Lumbar puncture was performed to do CSF analysis which showed CSF glucose, proteins, chlorides within normal limits and nil cells/organisms. MRI scan of the brain was performed which showed focal area of T2/FLAIR hyperintensity involving the splenium of corpus callosum, showing diffusion restriction on DW1, appearing dark on ADC. This picture was suggestive of cytotoxic lesions of corpus callosum (CLOCCs). EEG did not show any abnormal discharges. Complete fever work up was done for the patient out of which he reported positive for Weil Felix test (OX-19 ++++, OX-2 ++, OX K negative) suggestive of rickettsial infection. PCR could not be done due to financial constraints of the patient.

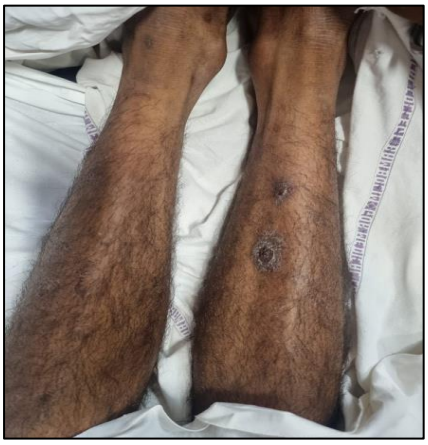

Figure 1: Eschar on the shin of the right leg.

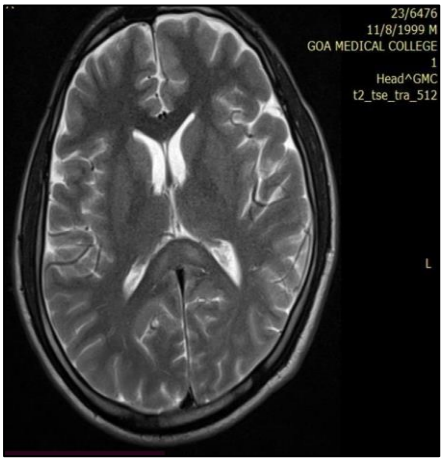

Figure 2: T2W axial image shows hyperintense signal within the splenium of corpus callosum.

Patient was initially empirically started on injectable ceftriaxone 2 gm BD. Later, injectable doxycycline 100mg BD added and was continued for 14 days. He was transfused 4 pints of RDP for thrombocytopenia. Over the next 5 days, patient's neurological status gradually improved. He was able to follow oral commands. Initially patient could comprehend but could not verbalize. Speech improved over time. He showed complete recovery in a span of 15 days and was discharged.

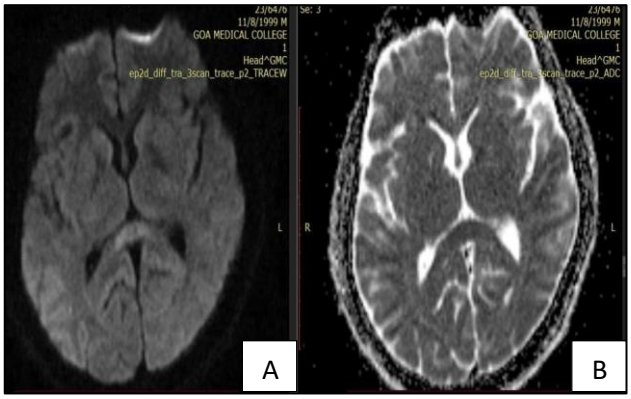

Figure 3 (A and B): DWI & ADC sequences of the same patient showing hyperintense signal with corresponding ADC values suggestive of restricted diffusion in the splenium of corpus callosum.

DISCUSSION

Rickettsia is a genus of small, rod-shaped round to pleomorphic obligate intracellular bacteria transmitted by vectors like lice, mites, ticks and fleas. Commonly encountered rickettsial pathogens include *R. Conorii* (Mediterranean spotted fever), *R. rickettsii* (Rocky Mountain spotted fever (RMSF), also known as Brazilian spotted fever); R. typhi (murine typhus); *Orientia tsutsugamushi* (scrub typhus); and *Anaplasma phagocytophilum* (anaplasmosis). Patient belonged to rural locality of Goa with many cases of rickettsiosis reported in recent times. Almost about 80% cases reported to CDC have missed data on eschars.[8] But, presence of an eschar evoked possibility of vector bite and consolidated our suspicion of the possible rickettsial infection.

Another interesting entity encountered in our case was the MRI finding of CLOCCS (cytotoxic lesions of corpus callosum). Markedly increased levels of cytokines and extracellular glutamate causes water to trap in callosal neurons and microglia presenting as cytotoxic edema.[10] On diffusion weighted MRI, these areas appear manifest as areas of low diffusion, particularly the splenium of corpus callosum. These lesions are commonly but not invariably reversible. Hence, also called as Reversible splenial lesions syndrome (RESLES).

A recent case of scrub typhus with pure alexia was reported to have similar MRI findings of CLOCCS. Ghosh R et al, suspected the cause to be unstructured/ functional

which the conventional MRI could not detect.[5] S. Houssem et al, also reported a case of Rickettsial encephalitis with CLOCCs in January 23. Their patient was a 22-year-old female who presented with history of headaches, global confusion and high-grade fever. Speech disturbances like dysarthria, verbal aspontaneity, stuttering, hypophonia and also aphasia have been reported previously in cases of corpus callosum injuries of varying etiologies.[12] Our patient had aphasia with no demonstrable structural abnormality on MRI in the primary language areas. Ishizaki M et al, had proposed in their article that the transcallosal diaschisis in the primary language cortex was responsible for the aphasia in their case of corpus callosal infarction.[13] Similar mechanism can be proposed in our case of corpus callosal edema. Hence, the global aphasia slowly improved completely as the edema resolved.

CONCLUSION

Rickettsial infections are the most underdiagnosed and underreported febrile illnesses requiring hospitalization. Most of the cases have good response to antibiotics like doxycycline. Varying presentations of these illnesses are now being reported. Hence, high index of suspicion and readily available diagnostic test can reduce the morbidity burden of these diseases.

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