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CASE REPORT

An Atypical Presentation of Rocky Mountain spotted fever (RMSF)- A Case Report

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ABSTRACT

Tick-borne illness has a highest incidence in South Central and South East U.S. The presence of Rickettsial diseases in India have been documented in Jammu and Kashmir, Himachal Pradesh, Uttaranchal, Rajasthan, Assam, West Bengal, Maharashtra, Kerala and Tamil Nadu [1],[2],[3],[4],[5],[6],[7],[8]. We report here, a case of a 70 year old man admitted with high grade fever and hypotensive and hypovolemic needed emergency intervention. Later, a Weil Felix test was done, followed by a demonstration of rise in titre. IgM ELISA and PCR confirmed the diagnosis of Rickettsiae. The patient was treated with Doxycycline and within 48 hours of treatment with Doxycycline, the patient was off ionotropes and was extubated. With gangrene of the extremities and superadded bacterial infection, the patient's WBC count had increased and the patient succumbed to death due to sepsis.

Key Words: RMSF, Rickettsia , Weil Felix test, Immunofluorescence, PCR

Key Message: High grade fever , hypotensive , hypovolemic , petechial rash, Weil felix test , IgM ELISA and PCR, RMSF

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differential diagnosis of fever. High case fatality rate in untreated individuals makes early recognition and treatment very essential. Our observation emphasizes that a rare presentation of fever with unusual signs and symptoms, if detected earlier, can be treated effectively.

Introduction

Rickettsiae rickettsii is a pleomorphic obligate intracellular gram negative bacillus which multiplies within the nuclei and the cytoplasm of host cells. The illness can be of apparent or severe form and death is reported to occur in 1% to 30% of untreated cases. Rocky mountain spotted fever should be considered in any individual who during the spring and summer has been in RMSF endemic areas and develops fever regardless of the presence of rash or the history of tick exposure [10]. Rickettsial fever does exist in our area, posing difficulty in the diagnosis and hence, it should be kept in mind as a

Case Report

A 70 year old man was admitted at Bhatia Hospital, Mumbai, with a history of high grade continuous fever for 2 days and an episode of general tonic clonic convulsions. On examination, the patient was found to be drowsy, arousable, febrile, hypotensive, hypovolemic and oliguric. The patient being tachypnoeic, had to be intubated and had to be put on a ventilator. He was given ionotropic support to maintain blood pressure and haemodialysis for anuria. The presence of petechial rash over the palm, which extended upto the wrist and lower extremities upto the ankle joint, made the

clinical diagnosis difficult. Further, with gangrene of the extremities and superadded bacterial infection, the patient's condition deteriorated. The history of tick bite could not be obtained as the patient had altered sensorium. Routine investigations being negative, made it difficult to arrive at a diagnosis for appropriate treatment.

Blood samples were sent for investigations: Investigations revealed a drop in Haemoglobin from 10.5g% to 7.8 g % on the 4th day.

WBC Count increased from 10,9400 to 13,010 cells / cumm. Random Blood sugar levels: 97mg/dL Platelets progressively decreased to 17 ,000 cells / cumm on the 7th day of illness, with the presence of giant platelets on peripheral smear examination. The creatinine level which was 4.6 mg/dL on day one, further increased to 6.8mg/dL on the 4th day. Hyponatraemia was 95mEq/L and hypocalcaemia was 5.3mg/dL. C 3 levels decreased to 81mg/dL.

Liver enzyme levels were elevated and the A/G ratio was reversed.

Tests for Malaria, IgM Leptospira, Dengue, HIV, HBsAg and Anti – HCV were negative. The blood culture showed no growth of organisms even after 1 week of aerobic incubation.

Emergency CT Scan revealed periventricular ischaemic changes.

Lumbar puncture was not done as bleeding parameters were derranged. Bleeding time: 2 min 30 sec. Clotting time was 3 min 15 seconds and APTT was 42.0 seconds.

The serum was tested for Weil Felix reaction. A titre of OX- 2 = 1 : 40 and OX – K 1 : 40 was observed. A four fold rise in titre was demonstrated in the sera with OX – K= 1 : 40 , OX -2 = 1 : 160 and OX – 19 = 1 : 160 after 10 days.

Further, blood samples were sent for Immunofluorescence - IgM ELISA to CMC Vellore, where it was reported as positive. To further confirm the diagnosis, the blood samples were sent to France for PCR. *Rickettsiae rickettsii* was identified as the causative agent by the PCR Weil Felix test and IgM ELISA and PCR confirmed the diagnosis of *Rickettsiae*. The immunofluorescence assay (IFA) is the gold standard technique and is used as a reference technique in most laboratories [11],[12] . Polymerase Chain Reaction is the confirmatory test, as reported elsewhere [15] .The patient was switched over to Doxycycline 100 mg b.d. and within 48 hours of treatment with Doxycycline, the patient was off ionotropes , he was extubated and had good urine output. The immediate response to treatment indirectly indicated Rickettsial infection, as Doxycycline is the drug of choice for rickettsial infection [9],[10] . IgM ELISA and PCR confirmed the diagnosis of *Rickettsiae* .

Discussion

Early clinical diagnosis is difficult, since the illness may have a gradual or an abrupt onset. The symptoms and signs may be unusual in timing or frequency and clinical appearance may vary depending on the age and location of the residence of the patients. The Weil Felix test was positive after 7 days of initial infection, during which time, the titre agglutinins in the patient's serum against *Proteus* strains OX- 2, OX 19 and OX –K were determined. The time from the onset of symptoms to death is less than 10 days and hence, a diagnosis of RMSF is seldom primarily made in survivors due to the atypical symptoms. Immunofluorescence assay (IFA) is the gold standard technique and is used as a reference technique in most laboratories.

The Weil Felix (WF) test is based on the detection of antibodies to various *proteus* species, which contains antigens with cross reacting epitopes to antigens from members of the genus *Rickettsia*. Even though the WF

agglutination test is not very sensitive, when positive, it is a rather specific test [12],[13],[14].

This report indicates a rare presentation of RMSF in which clinical conditions did improve initially after treatment with Doxycycline which is the antibiotic of choice as mentioned in other literature. But eventually, as a consequence of superadded bacterial infection due to multidrug resistant pseudomonas species in gangrenous areas of the extremities, the patient succumbed to death due to sepsis. Diagnosis should be largely be based on a high index of suspicion and careful clinical, laboratory and epidemiological evaluation supported by cost effective tests like WF, as the availability and the cost of Standard serological methods for Rickettsia are major problems in India. Empirical treatment should also be considered to reduce the high mortality and morbidity observed with the disease.

Conclusion

To conclude, recently, the disease has re-emerged in many areas of India. Routinely done serological investigations being negative, make it difficult to arrive at a diagnosis for appropriate treatment. Hence, Rickettsial infections should be kept in mind as a differential diagnosis of fever, as the case fatality rate in untreated individuals is very high. An efficient laboratory diagnosis can identify the causative agent even when there is no clinical suspicion. Successful cure from the disease can be achieved by following the proper antibiotic regimen. Immediate response to treatment indirectly indicated Rickettsial infection, as Doxycycline is the drug of choice -for the same [9],[10].

Abbreviations

RMSF - Rocky Mountain spotted fever

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