Case Report

Purpura fulminans secondary to indian tick typhus: a case report

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ABSTRACT

Purpura fulminans is a rare and commonly fatal syndrome that consists of hemorrhagic infarction of the skin and intravascular thrombosis. Purpura fulminans is commonly associated with streptococcal, staphylococcal and meningococcal infection. Indian tick typhus is a rare cause of purpura fulminans. One of the rare causes of purpura fulminans is Indian tick typhus, which is a type of rickettsial spotted fever caused by Rickettsia conorii and transmitted to humans by Rhipicephalus sanguineus (also called the dog tick). Clinical features of Indian tick typhus include fever, maculopapular rash that begins on the extremities with a centripetal spread, and constitutional symptoms including headache, malaise and conjunctival congestion. Severe cases can progress to multi-organ disease including pulmonary edema, meningoencephalitis, renal failure and cardiogenic shock can occur. Current report present a case of a 48 year old male patient who presented with features of Indian tick typhus which progressed to purpura fulminans and necrotizing fasciitis.

Keywords: Purpura fulminans, Rickettsia, Indian tick typhus, Retiform purpura, Gangrene

INTRODUCTION

A 48 year old male patient presented with fever, myalgia, vomiting, easy fatigability and blackish discoloration of bilateral second toes since 3 days. On initial examination, patient was febrile, BP was 130/80 mmHg, pulse rate was 88/minute, respiratory rate was 16/minute. Pallor, icterus, cyanosis, clubbing, edema and lymphadenopathy were absent. There was no history of pain abdomen, diarrhea, cough, sore throat, burning micturition or joint pains. There were no chills or rigors. He was a non-smoker and occasionally consumed alcohol. Cutaneous examination was unremarkable except for painless blackish discoloration of bilateral second toes suggesting impending gangrene (Figures 1 and 2).

CASE REPORT

Dorsalis pedis artery pulsations were feeble. Investigations showed hemoglobin was 17.4 g/dl, total leucocyte count was 16,510/mm³ (raised), platelet count was 0.88 lakhs/mm³ (low), serum Na was 133.0 mmol/l (low), urea 105 mg/dl (raised) and creatinine was 1.7 mg/dl (raised), liver enzyme; alkaline phosphatise: 237 U/l (raised) and serum bilirubin 6.6 mg/dl (raised), PT and APTT were normal, random blood sugar was 440 mg/dl (raised) and D-dimer was 2680 ng/ml (raised).

Figure 1: Gangrene of left second toe.
Patient was hospitalised and broad spectrum antibiotics piperacillin and tazobactam 2.25 grams TID were administered along with supportive measures.

The next day, the patient developed multiple petechiae over the abdomen, both upper limbs, palms and soles. Later that day, he developed altered sensorium followed by 2 episodes of seizures. He was immediately shifted to ICU wherein he was intubated and started on anti convulsants and other supportive measures. Five days later, he developed a generalized retiform purpuric rash over face, trunk, upper and lower limbs (Figure 3). Multiple hemorrhagic bullae developed over upper and lower limbs along with edema of both lower limbs. Surgery reference was sought and fasciotomy was performed on legs and feet (Figure 4).

Histopathology from the retiform purpuric rash on the right forearm showed areas congested vessels, perivascular mixed inflammatory infiltrate in the dermis along with hemorrhage and hemosiderin macrophages and occasional thrombosed vessels (Figure 5-6). Weil-Felix test showed a titer of 1:160 for OX-2; 1:20 for OX-19 and 1:40 for OX-K. Based on the findings of Weil-Felix test, patient was diagnosed with Indian tick typhus. Patient was started on injection doxycycline 100 mg twice daily. Topical applications including moisturizing lotion, fusidic acid cream and ointment containing benzyl nicotinate 2 mg and heparin 50 IU were given. After 5 days of treatment, there was marked reduction of skin lesions but the patient developed ventilator associated pneumonia caused by antibiotic resistant Klebsiella due to which general condition of the patient deteriorated and he succumbed as a result of sepsis.

DISCUSSION

Purpura fulminans first described by Guelliot in 1884, is a heterogeneous group of disorders characterized by...
rapidly progressive purpuric lesions that develop into extensive areas of skin necrosis, and peripheral gangrene.\textsuperscript{2,4} Three forms of purpura fulminans include acute infectious, neonatal and idiopathic purpura fulminans.\textsuperscript{4} The most common form is acute infectious purpura fulminans. This occurs most commonly due to acute bacterial sepsis including \textit{Staphylococcus aureus}, Streptococci and \textit{Hemophilus influenza}.\textsuperscript{4} Rickettsial infections have been rarely associated with purpura fulminans.\textsuperscript{2} Pathogenesis involves activation of complement and coagulation pathways by endotoxins and signaling by inflammatory cytokines or endothelial dysfunction and vasculitis caused by the antigen.\textsuperscript{2} Consequently there is acute transient decrease in protein C, protein S or antithrombin III levels.\textsuperscript{4} Complications of purpura fulminans include disseminated intravascular coagulation, end-organ damage, septicemic shock and necrotizing fascitis.\textsuperscript{4}

<table>
<thead>
<tr>
<th>Biogroup</th>
<th>Disease</th>
<th>Vector</th>
<th>Rickettsial species</th>
<th>Geographical distribution</th>
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</thead>
<tbody>
<tr>
<td>Typhus group</td>
<td>Endemic typhus</td>
<td>Rat flea (\textit{Xenopsylla cheopis}) Rarely cat flea (\textit{Cenocephalides felis})</td>
<td>\textit{Rickettsia typhi} \textit{Rickettsia felis}</td>
<td>Shimla, Kashmir, Jabalpur, Mumbai, Lucknow, Pune</td>
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<tr>
<td>Spotted fever group</td>
<td>Indian tick typhus</td>
<td>Tick (\textit{Rhipicephalus sanguineus})</td>
<td>\textit{Rickettsia conorii}</td>
<td>Nagpur, Jabalpur, Sagar, Pune, Lucknow, Bengaluru, Secunderabad</td>
</tr>
<tr>
<td></td>
<td>Flea-borne spotted fever</td>
<td>Rat flea (\textit{Ceratophyllus fasciatus})</td>
<td>\textit{Rickettsia species} (R14 strain)</td>
<td>Western Himalayan region; Himachal Pradesh</td>
</tr>
<tr>
<td>Scrub typhus (most common)</td>
<td>Scrub typhus</td>
<td>Mite (\textit{Trombiculidae})</td>
<td>\textit{Orientia tsutsugamushi}</td>
<td>Sub-Himalayan belt from Jammu to Nagaland; Sikkim, Darjeeling, Himachal Pradesh, Bihar, Rajasthan, Maharashtra; South India-Puducherry, Tamil Nadu, Kerala, Karnataka</td>
</tr>
</tbody>
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Rickettsial infections are caused by rickettsiae, which are small non-flagellate, gram negative, obligate intracellular parasites transmitted by arthropod vectors. Indian tick typhus, a zoonotic disease is a type of rickettsial spotted fever caused by \textit{Rickettsia conorii} and transmitted to humans by \textit{Rhipicephalus sanguineus} (dog tick).\textsuperscript{3,5}

In India, rickettsial infections have been reported from Himachal Pradesh, Maharashtra, Rajasthan, Assam, Jammu and Kashmir, Uttarakhand, Tamil Nadu, Karnataka, Kerala, Mizoram, and West Bengal (Table 1).\textsuperscript{6}

In Indian tick typhus, maculopapular rash begins on the extremities on third day of fever and has a centripetal spread. Eschar is rare and lymphadenopathy is usually absent. Constitutional symptoms include high grade fever, headache, malaise and conjunctival congestion.\textsuperscript{3} Complications like multi-organ disease including pulmonary edema, meningoencephalitis, renal failure and cardiogenic shock can occur.\textsuperscript{5} Weil-Felix test is the diagnostic test used to detect antibodies to various Proteus species antigens that cross-react with rickettsiae.\textsuperscript{6}

In current case patient initially presented with constitutional symptoms like fever and malaise. Since he was initially evaluated by a non-dermatologist and because of his dark complexion the maculo-papular rash may have been missed. Hence rickettsial fever may not have been considered in the initial work-up. During his hospital stay, he developed altered sensorium and was shifted to ICU. In the ICU, he developed skin rashes in the form of retiform purpura over his trunk and extremities. This is when we suspected rickettsial fever. Weil-Felix test was done and it was consistent with \textit{Rickettsia conorii}. He eventually developed purpura fulminans, necrotizing fascitis and other complications like ventilator associated pneumonia leading to sepsis and eventually succumbed to it.

**CONCLUSION**

Current case highlights the importance of keeping rickettsial infections in mind whenever we encounter cases of fever with rash, so that early diagnosis can be made and prompt treatment initiated, thereby reducing the morbidity and mortality in such cases.

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**Table 1: Geographic distribution of rickettsial infections in India.**\textsuperscript{6}
REFERENCES


