A pustule from Portugal

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A 42-year-old woman was admitted to hospital with a six-day history of fever as high as 40°C, chills, sweats and headache. She also complained of generalized weakness, sore throat and a nonproductive cough. These symptoms had been evaluated in the emergency room three days previously, at which time a small cutaneous lesion on the upper medial portion of the left breast was noted. She was given a prescription for cefuroxime 500 mg twice daily, which she took, but she did not improve. She returned on the day of admission because of a new nonpruritic rash over her entire body, including the palms of her hands and the soles of her feet.

The patient was a schoolteacher and had no significant past medical history. She had returned from a one-month vacation in Portugal one day before the onset of her symptoms. No family members were ill. She had stayed mainly in the countryside and had gone hiking in wooded areas.

On examination, she appeared to be ill with a temperature of 38.9°C, a pulse of 145 beats/min, a blood pressure of 110/50 mmHg and a respiratory rate of 18 breaths/min. There was a slight conjunctival injection. The cardiorespiratory

examination was normal and no organomegaly or adenopathy was present. Her body was covered with a nonblanching erythematous and ecchymotic maculopapular rash that involved the palms and soles. A small (1 to 2 mm) crusted cutaneous ulceration was present on the left breast.

Relevant laboratory data included a hemoglobin level of 118 g/L, leukocyte count of 6.3×10^9 /L, platelet count of 124×10^9 /L, alkaline phosphatase level of 341 IU/L (three times the normal level), alanine aminotransferase level of 350 IU/L (nine times the normal level), aspartate aminotransferase level of 385 IU/L (nine times the normal level), gamma-glutamyl transpeptidase level of 208 (four times the normal level), and a normal chest radiograph and abdominal ultrasound. She continued to have daily fevers up to 39°C and her leukocyte count rose to 16.8×10^9 /L. Serological tests for hepatitis A immunoglobulin M (IgM), hepatitis B surface antigen, rubella IgM and measles IgM were all negative.

What is your diagnosis and what therapy would you initiate?

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DIAGNOSIS

On the strong suspicion of a rickettsial disease, the patient was started on gatifloxacin pending results of serological testing. There was some decrease in the rash and general improvement, but the fever persisted for another 48 h and the patient was then switched to oral doxycycline 100 mg twice daily. Within 24 h of the switch, she became afebrile and the rash resolved. The patient was discharged from hospital and all laboratory parameters had returned to normal on a follow-up visit one week later. Serological testing for rickettsial disease was performed by Dorothy McColl at the Canadian Science Centre for Human and Animal Health (Winnipeg, Manitoba) and showed an initial titre of less than 1:32, with a convalescent titre of 1:512 to *Rickettsia rickettsii*. The causative agent of Mediterranean spotted fever is *Rickettsia conori* and specific testing for this organism is unavailable in Canada. However, there is serological cross-reactivity among members of the spotted fever group. It is our belief that the patient had contracted Mediterranean spotted fever (Boutonneuse fever) in Portugal.

DISCUSSION

Mediterranean spotted fever is acquired through the bite of the dog tick, *Rhipicephalus sanguineus*. The incubation period is five to seven days after an often ignored tick bite. Symptoms usually include fever, headache, myalgias, malaise, gastrointestinal disturbance and rash. There is endothelial invasion by the

organism, leading to vasculitis and tissue necrosis. This is manifested at the tick bite site by the formation of an eschar, the 'tache noire'. Complications of infection include polyneuropathy (1), renal failure, thrombocytopenia and even death (2). The diagnosis can be made by combining the clinical and epidemiological information and performing rickettsial serological testing on acute and convalescent samples. In some centres, blood and/or skin biopsy specimens have been cultured in cell lines (3) or inoculated into guinea pigs. The treatment of choice is doxycycline but quinolones and the newer macrolides have also been used successfully (4,5).

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