Journal of the Postgraduate Institute of Medicine 2018; 5 (1): E61 1-6
http://doi.org/10.4038/jpgim.8167

Case Report

A rare case of spotted fever group of *Rickettsia* infection with fever, facial oedema and fern leaf necrosis of the skin

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Keywords: Spotted Fever, *Rickettsia* Infections, Tropical Diseases, Bacterial Infections, reemerging infections

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Received: October 2017, Accepted revised version January 2018, Published: March 2018
Competing Interests: Authors have declared that no competing interests exist

Background

Rickettsial infections, also known as typhus or rickettsioses, are important causes of febrile illness, especially in the Asia Pacific region [1]. They are caused by obligate intracellular coccobacilli of the Rickettsiaciae family and infect humans through arthropod vectors [1]. Typhus is an important reemerging infection in Sri Lanka [2]. A high degree of clinical suspicion and early institution of treatment is mandatory to prevent case fatality due to multi organ involvement [3].

Case Presentation

A 49 year old lady from Handawalpitiya, a rural mountainous region of Sri Lanka, was transferred to Teaching Hospital Kandy for further management of fever and thrombocytopaenia. She was a previously healthy woman. She had fever with chills of six days duration which was associated with generalized myalgia, arthralgia and headache. She had no contact history of fever or any exposure to contaminated water. She was ill looking with periorbital and perioral oedema (Figure 1). She also had mild ankle oedema. There was no pallor or icterus. She was tachypnoeic with a respiratory rate of 30 per minute and had a few right basal crepitations. Patient was haemodynamically stable on admission with normal jugular venous pressure. She had tender hepatomegaly, but no splenomegaly. Her neurological examination was normal.

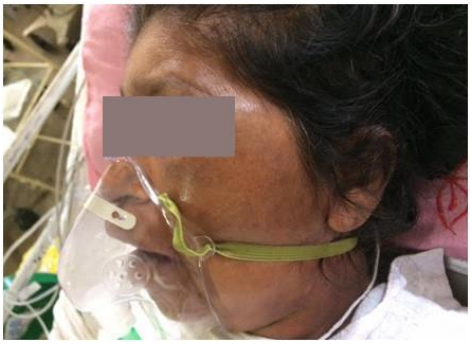

Figure 1: Facial oedema on Day 6 of the disease.

On the 2nd day of admission she went into a confusional state and developed a generalized ecchymotic eruption mainly involving the trunk and extremities (Figure 2, Figure 3).

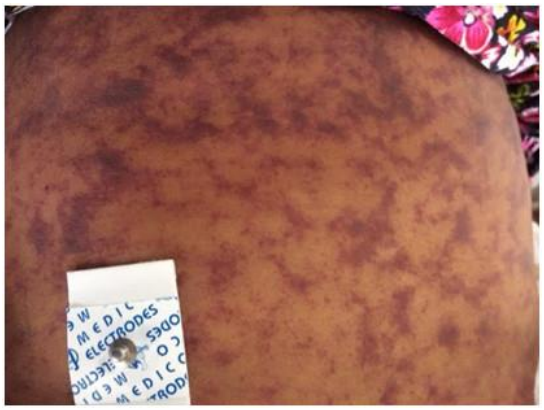

Figure 2: Fern leaf necrosis of the skin.

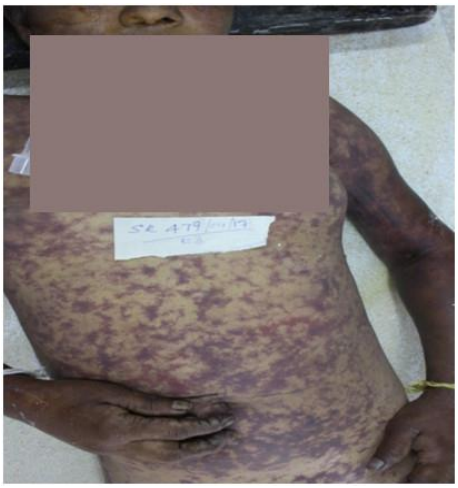

Figure 3: Fern leaf necrosis of the skin.

She had no neck stiffness. Fundi were normal and deep tendon reflexes were normal with bilateral down going plantar responses. Her blood pressure was 110/70mmHg with tachycardia and tachypnea with a respiratory rate of 40 per minute. She could maintain normal oxygen saturation. Later on the same day she was transferred to the intensive care unit as her blood pressure started dropping and she needed inotropic and ventilator support.

Table 1: Investigation summary

Investigation	D1	D2	D3		Investigation	D1	D2	D3
White Blood Cells(10^9/L)	4.4	6.7	7.2		ESR (mm/1st hour)	10		
Neutrophils (%)	86	92	91		CRP (mg/L)	221		
Lymphocytes	6.2	6.1	6.8		CPK(U/L)	1585		
Platelets (10^9/L)	13	10	07		Serum Na^+(mmol/L)	139		
Haemoglobin (g/L)	11.2	11.7	11.2		Serum K^+ (mmol/L)	3.7		
Haematocrit (%)	32	34			Serum Creatinine (mg/L)	0.5	1.2	
					Urine Full Report			
ALT (U/L)	144	135			Proteins	++		
AST (U/L)	286	319			Red Cells	6-8		
ALP (U/L)	220				Pus Cells	2-5		
GGT (U/L)	491				Granular Casts	+		
Serum Albumin (g/dL)					PT/INR	Normal		
Total Protein (g/dL)					APTT	Normal		

ALT= Alanine amino transferase; AST=Aspartate amino transferase; ALP=Alkaline phosphatase; GGT=Gamma glutamyl transferase: ESR=Erythrocyte Sedimentation Rate; CRP=C-reactive protein; CPK=Creatine Phosphokinase; S. Na^+=Serum Sodium; S. K^+=Serum Potassium; PT=Thrombin Time; INR= International Normalised Ratio; APPT=Activated Partial Thromboplastin Time

Her blood picture revealed a reactive film with marked thrombocytopaenia. Blood cultures and urine cultures were negative. Dengue antibodies were negative on day 7 of the disease (2nd day of admission).

Ultra sound scan of the abdomen and chest revealed mild hepatomegaly with oedema of the gall bladder wall and no pleural effusion or free fluid in the abdomen. Chest X-ray showed a small inflammatory shadow in the right lower zone.

Due to the high prevalence of dengue in the region, she was initially managed as dengue haemorrhagic fever complicated by fluid over-load and secondary sepsis. With the appearance of the rash, clinical suspicion shifted towards a rickettsial infection and she was started on doxycycline with IV dexamethasone. However, in-spite of treatment, she deteriorated and passed away on the 3rd day after the admission. Post mortem revealed congested organs with haemorrhagic manifestations. (Figure 4)

Her blood was sent for typhus serology and became positive for infection with the spotted fever group of *Rickettsia*.

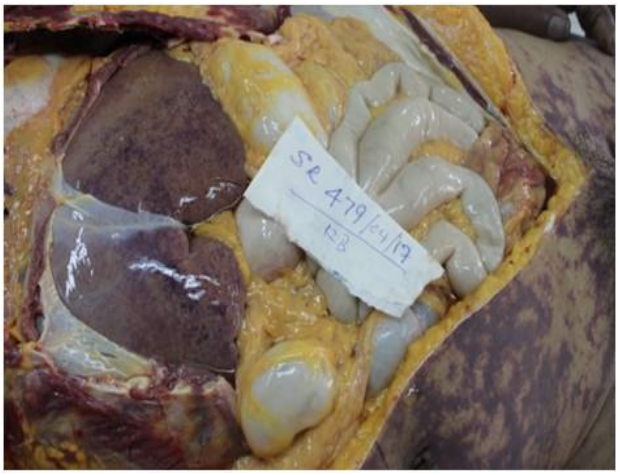

Figure 4: Post mortem findings of haemorrhagic manifestations

Discussion and Conclusion

Rickettsial infections include diseases caused by bacteria of the Genus *Rickettsia*, which include the spotted fever group and the typhus group and scrub typhus caused by *Orientia tsutsugamushi*. Rickettsia parasitize the endothelial cells resulting in a multi systemic vasculitis leading to a wide array of clinical manifestations.

Clinical manifestations of rickettsial disease range from subclinical infection to multi organ failure. Onset of the illness is abrupt with sudden onset of fever and headache and a rash that appears on day 4 to 7 of the illness. This can be macular, maculopapular or petechial, starting from the trunk and spreading to the periphery. The extent and severity of the rash is highly variable and in the more severe forms of illness, the rash may be necrotic with a fern leaf appearance with serrated margins known as fern leaf necrosis. Eschar, the site of the tick bite, is present in 60% of cases of the scrub form of typhus. Also, in general, the facial and leg skin of the patient seems slightly oedematous particularly in elderly patients. Characteristic skin lesions play a key role in the clinical

diagnosis of the spotted fever group of rickettsiosis [4]. Diverse neurological manifestations are also reported [2]. Disease spectrum varies, mainly depending on the rickettsial species [4].

The diagnosis of rickettsiosis is often made clinically, as the specific diagnostic investigations are available in only a few laboratories in the world. Diagnostic methods include serological tests, such as indirect immunofluorescence assay or enzyme immune assay or PCR based methods to identify species. Because of the lack of diagnostic testing in endemic areas, as well as non-specificity of symptoms, many cases are undiagnosed, or rather misdiagnosed as other infectious diseases like leptospirosis, meningococcaemia or even dengue [5,3]. Also, serology is neither sensitive nor informative during the acute phase of the illness [6]. Untreated rickettsiosis, especially of the spotted fever group, has high fatality rates and timely treatment decreases mortality and morbidity [7,8,9,10]. Doxycycline and chloramphenicol are the major anti rickettsial agents used to treat typhus. Doxycycline resistant typhus has been reported in Thailand and azithromycin and rifampicin have been shown to be effective in these cases in small trials [9,10,11]. Intravenous hydrocortisone, in addition to antibiotics, has shown to be effective in severe disease with evidence of multi system vasculitis [12].

Hospital-based studies, carried out in 2002 to 2008, described predominance of spotted fever group rickettsia in the hilly Central Province of Sri Lanka [12, 13]. In contrast to scrub typhus and murine typhus, the incidence of infection due to the spotted fever group appears to be increasing. Clinically, patients with spotted fever have severe illness compared with other rickettsial infections [2]. All the patients with confirmed spotted fever have skin rash of varying severity. Some may have “fern leaf necrotic skin rash” as well as diverse neurological manifestations [2]. A necrotic rash, elevated liver enzymes, thrombocytopaenia and high ESR are warning signs of life threatening multi organ dysfunction [14]. The most recognized vector for spotted fever is the hard tick that parasitizes every class of vertebrae in almost every region of the world [2,14,13].

During the last two decades, novel rickettsia have been isolated and characterized. The emergence or re-emergence of rickettsial infections is occurring in many regions of world. Thus, clinicians should have a high degree of suspicion and be vigilant to detect novel clinical manifestations [2]. Thus, the empiric use of anti-rickettsial antibiotics is justified in patients with undiagnosed fever who have no clinical features or investigation results suggesting another aetiology in a setting where rickettsial infections are endemic or re-emerging [14]. If the illness is rickettsial in origin, the patient may have a dramatic clinical response, even in the presence of serious complications. Thus mortality and morbidity may be greatly reduced [14]. However, establishment of proper diagnostic facilities is urgently required to improve early diagnosis and to reduce mortality and morbidity. It is also important to understand the varying clinical manifestations and the changing epidemiology of this disease.

Declarations

Consent for publication: Written informed consent was taken from the family of the patient for publication of this case report.

Availability of data and material: Investigations and patient records available on request.

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