

thalassaemia intermedia. Independent mutation is the most likely explanation for the occurrence of Hb Hofu in individuals of Japanese, Spanish, Indian and Sri Lankan origin [1-5].

Routine baseline thalassemia screening involves automated FBC to evaluate red cell indices. This does not detect any abnormality in the heterozygous state of Hb Hofu. Hb Hofu was detected by Hb HPLC but can be overlooked due to a close association with Hb A0. Hence, partner screening of a known carrier, especially with beta thalassaemia trait, should include haemoglobin HPLC with careful interpretation.

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Thrombotic microangiopathy following Russell's viper (*Daboia russelii*) envenoming in Sri Lanka: a case report

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Introduction

Thrombotic microangiopathy (TMA), though uncommon, is a recognized complication of snake envenoming [1]. Characteristically, it comprises of the triad of microangiopathic haemolytic anaemia, thrombocytopenia and acute renal failure [1]. Association of TMA with hump-nosed viper (*Hypnale* species) envenoming has been described recently in Sri Lanka [2]. However, association of TMA with Russell's viper (*Daboia russelii*) envenoming had not been clearly defined except in a publication in 1975 in India which described manifestations of TMA in 5 patients [3]. It appears that TMA is a rare manifestation of Russell's viper bite, despite the fact that its bites are common in Sri Lanka [4]. We present a patient with severe anaemia, thrombocytopenia and mild acute renal failure after Russell's viper bite in Kandy district suggestive of TMA.

Case report

A 43 year-old woman was working in a paddy field in

a remote village in the Central Province. At about 11 am her foot was bitten by a Russell's viper. With a vigorous shake the snake was dislodged and she ran to find help. But she stumbled and fell down on her face and stuck the forehead. Then she started to bleed heavily from the nose and fainted within a few minutes. After regaining consciousness, she managed to wave her hands and got the attention of another farmer. She was brought to a local hospital within 30 minutes. On admission to the hospital she had ophthalmoplegia and passed red coloured urine suggestive of haematuria. The bleeding scalp laceration was managed with tight bandaging. She was given 10 vials of Indian polyvalent antivenom and transferred to a tertiary care hospital where she underwent CT scan of the brain which was reported normal. However, over the next two hours her blood pressure dropped and therefore she was sent to the intensive care unit of the Teaching Hospital, Peradeniya. Upon resuscitation with intravenous fluid and hydrocortisone her blood pressure increased to 110/70 mmHg. The radial pulse rate was 98 min. She had bilateral ptosis and external ophthalmoplegia, but respiration was normal. Fang marks were present over

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Table 1.

	Day 1	Day 2	Day 3	Day 4	Day 5	Day 5	Day 6	Day 8
WBC count ($\times 10^3/\text{mm}^3$)	3.7	27.0	16.0	23.6	17.7	15.4	20.7	9.9
Neutrophils %	57.0	90.0	80.0	93.1	84.8	84.8	72.6	71.8
Lymphocytes %	40.0	10.0	20.0	5.6	12.0	12.8	13.2	17.3
Hb g/dl	10.0	9.5	7.1	9.8	11.3	11.2	11.2	11.4
PCV %	29.0	26.0	22.0	29.5	33.2	34.5	34.1	35.9
Platelet count ($\times 10^3/\text{mm}^3$)	236	70	40	10	21	20	29	101
Creatinine $\mu\text{mol/l}$			168	198		80		66
ALT U/l				87				

WBC – white blood cells, N – neutrophils, L – lymphocytes, Hb – haemoglobin, PCV – packed cell volume, Plt – platelet count), Creatinine – serum creatinine, ALT – alanine transaminase

the dorsal aspect of right foot about 2 cm distal to the ankle joint from which blood was oozing. As whole blood clotting time (20WBCT) was more than 20 minutes a further 15 vials of antivenom was infused over one hour. Thereafter, the bleeding stopped and repeat 20WBCT was normal. However, regular clinical monitoring showed anaemia which was confirmed by haemoglobin levels.

On the third day the patient was tachypnoeic and showed conjunctival pallor. The WBC was $16,000/\text{mm}^3$, haemoglobin level was 7.1g/ dl, and platelets were $40,000/\text{mm}^3$. The serum creatinine on day 3 was $168\ \mu\text{mol/l}$ (Table 1). On the 4th day platelet count dropped to $10,000/\text{mm}^3$ and the blood picture done before blood transfusion showed normochromic, normocytic, polychromatic red cells and fragmented red cells suggestive of microangiopathic haemolytic anaemia. Three pints of blood and 8 units of platelet concentrate were transfused, and her condition improved with the treatment. There was no reduction of urine output; however, a transient rise of serum creatinine ($198\ \mu\text{mol/l}$) occurred. On discharge her haemoglobin level had increased to 11.4 g/dl and platelet count to $101,000/\text{mm}^3$ (Table 1).

Discussion

After initial treatment with antivenom, the patient developed microangiopathic haemolytic anaemia on the 3rd day (Table 1). Thrombocytopenia manifested from day 2 to 6 and renal dysfunction was observed on the 4th day suggestive of TMA.

The concept of venom-induced consumption coagulopathy (VICC) has replaced the former description of disseminated intravascular coagulation (DIC) as the commonest coagulation pathology in envenoming [5]. In VICC, activation of coagulation pathway occurs due to snake toxins such as thrombin-like enzymes, prothombin

activators and factor X activators [5]. It is argued that when TMA combines with VICC, the manifestations are similar to the manifestations of DIC [5]. TMA is a new concept described recently. The pathogenesis is related to ADAMTS13 which is a metalloprotease enzyme that cleave the large von Willebrand factor which inhibits spontaneous activation of platelet aggregation [5]. When ADAMTS13 activity is normal, plasmapheresis may not be beneficial in snake envenoming causing TMA [5]. In conclusion, we highlight TMA as a potential haematological problem after Russell's viper bite in Sri Lanka. The diagnosis of TMA may have been overlooked in the past and further research is recommended.

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