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Cutaneous involvement in haemolytic uraemic syndrome

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Case

A 35-year-old woman was admitted to our hospital with severe hypertension, oedema and renal failure. Twelve months earlier, she had a pregnancy, which was complicated by preeclampsia. A successful Caesarean was performed in the 29th week of gestation (Figures 1 and 2). After the birth, she remained with poorly controlled hypertension and proteinuria exceeding 1 g/day. Six months later, she began to have fatigue, headaches, severe hypertension and facial and pedal oedema. There was no fever, neurologic abnormalities or diarrhoeal prodrome. She was pale and hydrated and blood pressure was 151/103 mmHg. Laboratory screening revealed haemoglobin 4.65 mmol/L (7.5 g/dL), urea 182 mg/dL, creatinine 65 mmol/L (6.46 mg/dL), platelets 46 × 109/L, increased reticulocyte count and schistocytes in peripheral blood. Proteinuria value was 4.9 g/24 h and urinary sediment revealed 5–10 red blood cells/HPF. The remaining study showed lactic dehydrogenase 1669 U/L, normal liver enzymes, total bilirubin 14 mg/L, direct bilirubin 4.4 mg/L, haptoglobin <0.2 mg/dL and negative Coombs test. All immunologic tests and viral screening were normal. A diagnosis of atypical haemolytic uraemic syndrome (aHUS) was established, and she started daily plasmapheresis with haemodialysis. After 6 months, haematological parameters normalized but she never recovered renal function and remained on dialysis. Seven months later, she had a relapse of the aHUS and needed plasmapheresis during 6 months. After 4 months, she complained of three painful leg skin lesions which progressed to ulcers, with some necrotic areas of the base and edges. One month later, she again developed uncontrolled hypertension and relapse of the haemolytic activity. Immunologic studies including antiphospholipid antibodies remained normal. Skin biopsy showed fibrin thrombi in the small blood vessels, with discrete inflammatory infiltrate of lymphocytes and plasma cells. A new relapse of aHUS was then diagnosed and she was once again treated with plasmapheresis, with resolution of haematological and cutaneous disorders after 11 weeks. aHUS is a rare, systemic, life-threatening disease [1, 2], caused by dysregulation of the alternative pathway of complement [3]. Although being extremely rare, we should be mindful of skin involvement in aHUS and establish this association in a timely manner which would lead to an early diagnosis and correct treatment, avoiding wasted time and costs [2].

References

1. Noris M, Remuzzi G. Atypical hemolytic-uremic syndrome. *N Engl J Med* 2009; 361: 1676–1687
2. Siegler RL. Spectrum of extrarenal involvement in postdiarrheal hemolytic uremic syndrome. *J Pediatr* 1994; 125: 511–518
3. Loirat C, Noris M, Fremeaus-Bacchi V. Complement and the atypical hemolytic uremic syndrome in children. *Pediatr Nephrol* 2008; 23: 1957–1972

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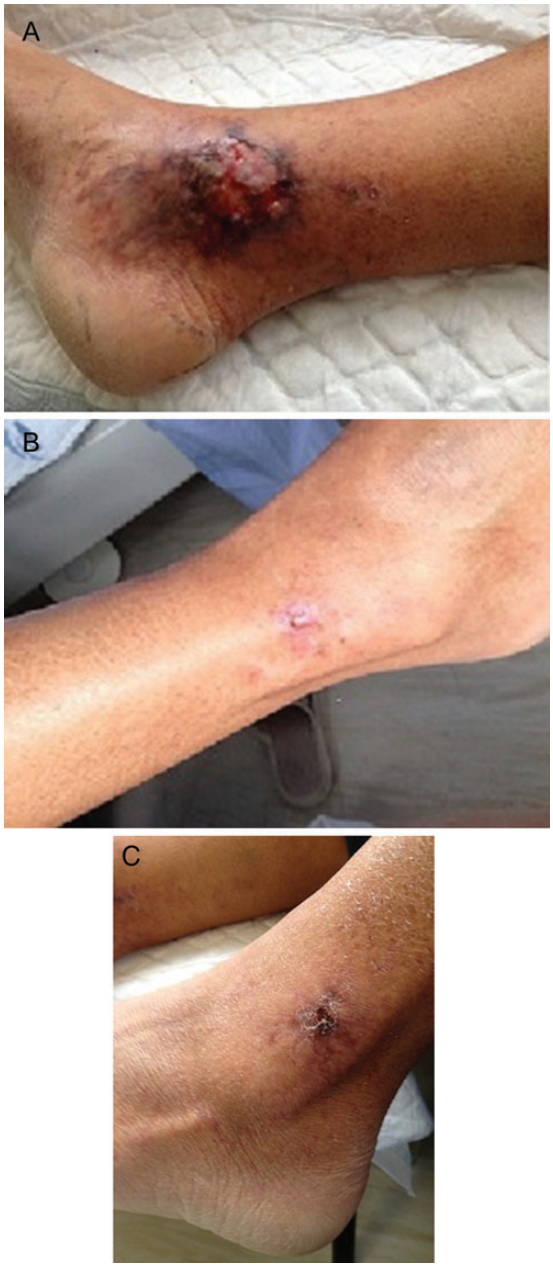

Fig. 1. (A) Leg skin lesions with purplish colour before plasmapheresis. (B) Leg skin lesions with purplish colour before plasmapheresis. (C) Leg skin lesions with purplish colour before plasmapheresis.

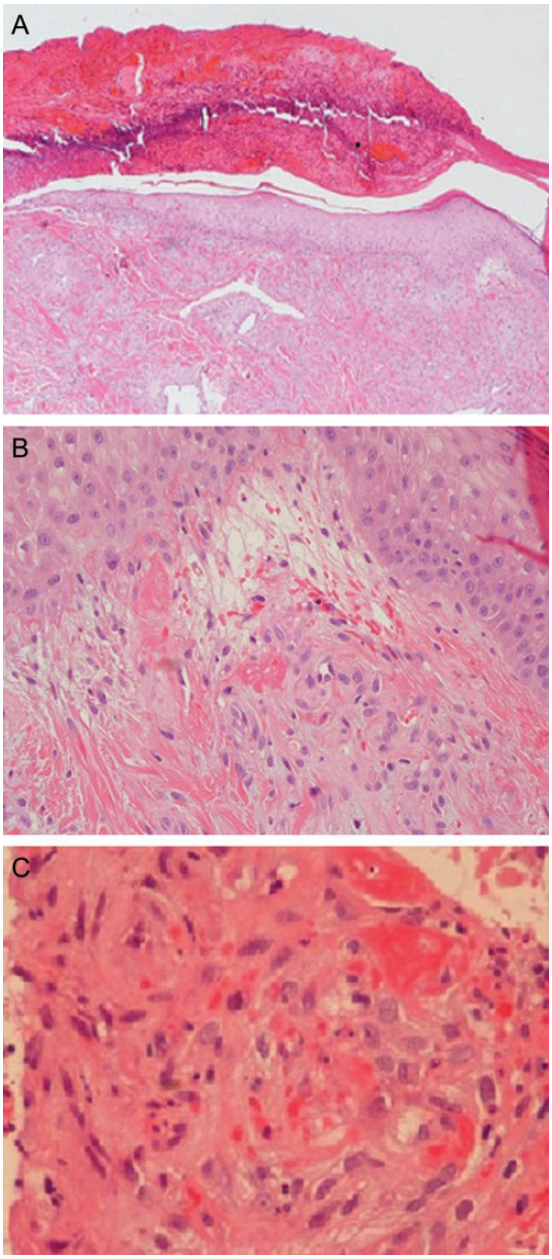

Fig. 2. (A) Skin biopsy showed fibrin thrombi in the small blood vessels, with discrete inflammatory infiltrate of lymphocytes and plasma cells. (B) Skin biopsy showed fibrin thrombi in the small blood vessels, with discrete inflammatory infiltrate of lymphocytes and plasma cells. (C) Skin biopsy showed fibrin thrombi in the small blood vessels, with discrete inflammatory infiltrate of lymphocytes and plasma cells.