

To the Editors:

## A case of paroxysmal cold haemoglobinuria following chickenpox

Paroxysmal Cold Haemoglobinuria (PCH) is a rare type of cold haemolytic anaemia (HA), characterized by intravascular haemolysis due to an IgG antibody named after Donath Landsteiner. PCH is usually associated with viral infections, but association with varicella zoster infection (VZI) is rare in adults. We present the second such reported adult case, which is the first in Sri Lanka [1].

A 17 year old, previously healthy girl, developed vomiting, fever and faintness, and two bouts of dark red urine suggestive of haemoglobinuria. She had features of recent VZI, with healing vesicles. She was pale, jaundiced, and had features of heart failure. She had marginal hepatomegaly on abdominal examination.

Investigations showed a very low Hb of 2.3 g/dl, platelet count of  $3.89 \times 10^9/l$ , and white cell count of  $11.8 \times 10^9/l$  (neutrophils 85%). Blood picture showed cold type HA, but failed to show any erythro-phagocytic neutrophils. She had a high ESR (125mm/hr) and high ferritin (312 nmol/l [ref. 0.27 - 0.337 nmol/l]) levels. Serum iron was 178  $\mu\text{mol/l}$ , TIBC was 240  $\mu\text{mol/l}$  and transferrin saturation was 74%. Liver function tests showed increased serum bilirubin levels (total 25.6 mmol/l, direct 5.2 mmol/l) with normal enzyme levels. She had normal abdominal ultra sonography. Renal function tests, urine analysis and culture were normal. Direct Coomb's test was weakly positive with poly-specific anti-human globulin, and the indirect test was negative. Cold agglutinins were negative after screening at 4 °C against adult (I), cord (i) and autologous erythrocytes. Donath-Landsteiner antibody was positive, confirming the diagnosis. However, anti-P antigen specificity was not tested. Mycoplasma antibodies, HIV screening, VDRL, hepatitis B antibodies, Rheumatoid Factor and Anti Nuclear Antibodies were negative. G6PD assay, Epstein-Barr serology and osmotic fragility were not performed.

She was treated with high dose prednisolone (1mg / kg / day), with six packs of leucodepleted P antigen positive red cell concentrates (LDRCC) (cross matched at 37 °C), folate supplementation and was nursed warm. After one week, she was discharged with a Hb of 9 g/dl, with no further evidence of haemolysis. Three months later, she was asymptomatic.

PCH was first described as a chronic relapsing haemolytic anemia, but this picture is rare now. The acute non-relapsing form of PCH has been documented to occur in adults as isolated cases, associated with illnesses such as infectious mononucleosis, *Mycoplasma pneumoniae* and *Klebsiella pneumoniae* infections, non-Hodgkin lymphoma [1], myelo-dysplastic syndromes [2], myeloproliferative syndromes [3], and small cell carcinoma [4].

The management of the condition was based on isolated case reports. Steroid therapy has generally not been found to be beneficial, but may be useful in atypical presentations (i.e. Evan syndrome) [1,4]. Rituximab and azathioprine have been shown to be beneficial in refractory cases [5,6].

### References

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