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<th><strong>Title</strong></th>
<th>Transfusion-refractory anaemia in liver cirrhosis</th>
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Transfusion-refractory anaemia in liver cirrhosis

CLINICAL PRESENTATION
A 33-year-old-man with chronic alcoholism presented with anaemia. Investigations showed haemoglobin 7.5 g/dl (reticulocytes: 11.2%), leucocytes 4.7×10⁹/l, platelets 61×10⁹/l, direct bilirubin 221 (reference <6) μmol/l, aspartate transaminase 109 (reference 15–38) U/l, alanine transaminase 38 (reference 8–58) U/l, γ-glutamyltransferase 31 (reference 11–62) U/l, lactate dehydrogenase 319 (reference 118–221) U/l, haptoglobin <0.06 (reference 0.16–1.97) g/l, methaemalbumin 0.18 (reference <0.1) mg/dl and a negative direct antiglobulin test. The iron status, cholesterol and triglyceride profiles were unremarkable. On referral, physical examination showed pallor, jaundice, hepatomegaly (3 cm), splenomegaly (10 cm) and ascites. Seven units of blood were transfused without improvement of his anaemia. Trans-jugular liver biopsy confirmed alcoholic cirrhosis. The peripheral blood film showed dysmorphic red cells (fig 1A, arrows).

QUESTION
What was the diagnosis?
See page 114 for answers
ANSWER

From the question on page 5

There was significant echinocytosis, confirmed by scanning electron microscopy (SEM) (fig 1A, arrows). Serial blood films and SEM after a four-unit transfusion showed a decrease followed by progressive increase in echinocytes (fig 1B,C), indicating that transfused red cells also became echinocytes. The diagnosis was haemolytic anaemia due to echinocytosis secondary to cirrhosis.

Echinocytosis might be found in cirrhosis of different aetiologies, where abnormal plasma high-density lipoproteins (HDLs) are present, owing to decreased hepatic clearance. Abnormal HDL incorporation into the red cell membrane perturbs its structure, leading to echinocytosis. An intrinsic red cell metabolic defect is not involved. Hence, transfused red cells also undergo echinocytic transformation. Echinocytes are poorly deformable and are destroyed during microcirculation filtration.

This condition is different from alcoholic hepatitis-induced Zieve syndrome, with hyperlipidaemia and haemolytic anaemia. Acute alcoholic intoxication damages the red cell metabolism, leading to an acquired pyruvate kinase deficiency and plasma membrane oxidation, resulting in haemolysis.

REFERENCES
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