



Title	Transfusion-refractory anaemia in liver cirrhosis
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Citation	Gut, 2010, v. 59 n. 1, p. 5+114
Issued Date	2010
URL	http://hdl.handle.net/10722/129502
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Editor's quiz: GI snapshot

Emad El-Omar, *editor*

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CLINICAL PRESENTATION

A 33-year-old-man with chronic alcoholism presented with anaemia. Investigations showed haemoglobin 7.5 g/dl (reticulocytes: 11.2%), leucocytes $4.7 \times 10^9/l$, platelets $61 \times 10^9/l$, direct bilirubin 221 (reference <6) $\mu\text{mol/l}$, 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

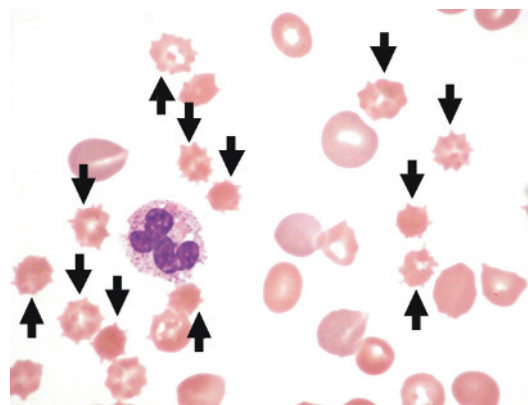


Figure 1 A peripheral blood film, showing numerous dysmorphic red blood cells (arrows) (Wright stain, original magnification $\times 1000$).

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Competing interests: None.

Patient consent: Obtained.

Provenance and peer review: Not commissioned; externally peer reviewed.

Gut 2010;**59**:5. doi:10.1136/gut.2009.182832

Editor's quiz: GI snapshot

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,¹ where abnormal plasma high-density lipoproteins (HDLs) are present,¹ 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

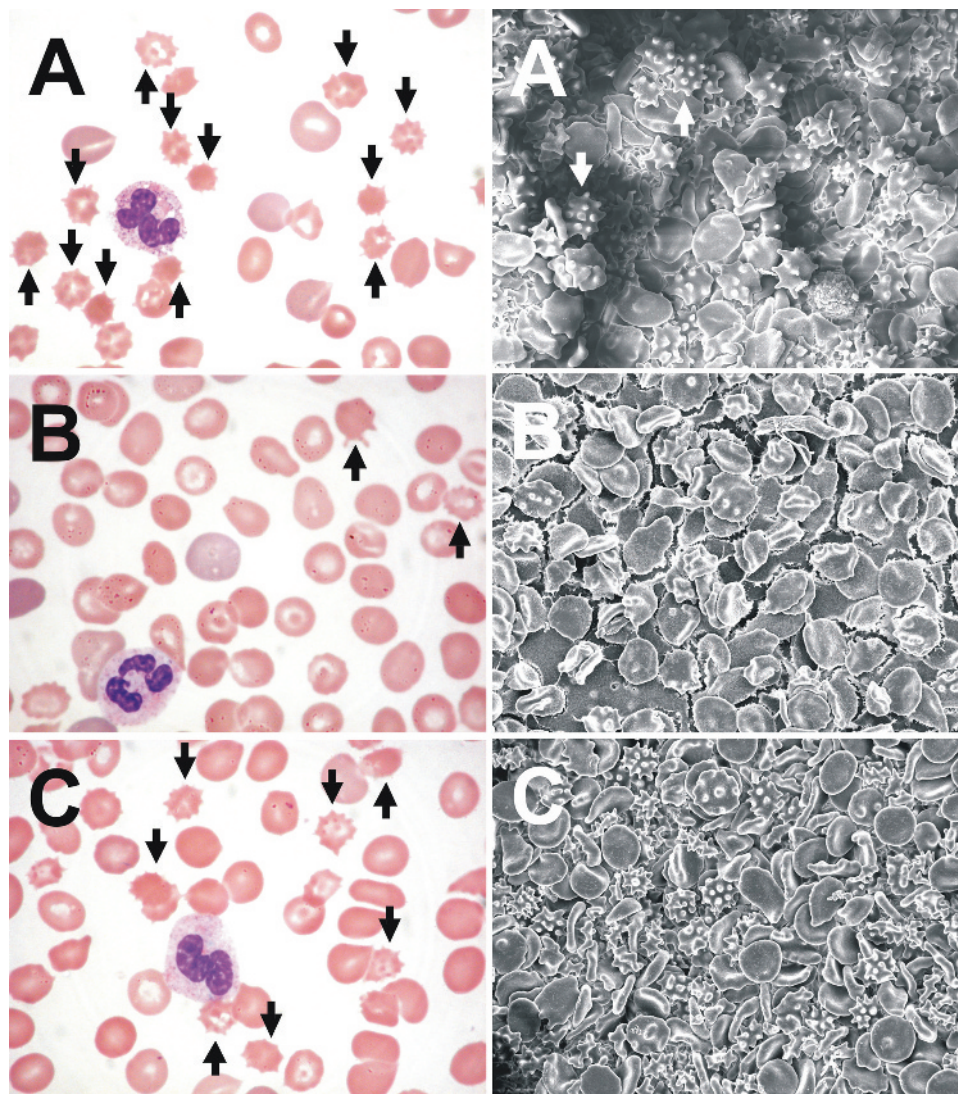
transformation. Echinocytes are poorly deformable and are destroyed during microcirculation filtration.²

This condition is different from alcoholic hepatitis-induced Zieve syndrome, with hyperlipidaemia and haemolytic anaemia.³ Acute alcoholic intoxication damages the red cell metabolism, leading to an acquired pyruvate kinase deficiency and plasma membrane oxidation, resulting in haemolysis.⁴

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Figure 2 A peripheral blood film, showing numerous dysmorphic red blood cells (arrows) (Wright stain, original magnification $\times 1000$). (A) Peripheral blood film showing significant echinocytosis (arrows). This was confirmed by scanning electron microscopy (SEM), which showed numerous echinocytes (burr cells), characterised by membrane crenation with numerous small spicules (arrows) (Latin, *echinus*, meaning sea urchin). (B) Peripheral blood film 12 h after blood transfusion, showing only occasional echinocytes (arrows). This was confirmed with SEM. (C) Peripheral blood film 48 h after blood transfusion, showing a significant increase in the number of echinocytes (arrows), indicating that the transfused red cells underwent echinocytic transformation. This was confirmed with SEM.





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Gut 2010 59: 5

doi: 10.1136/gut.2009.182832

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