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PHOTO QUIZ

A diagnosis on the basis of a blood smear

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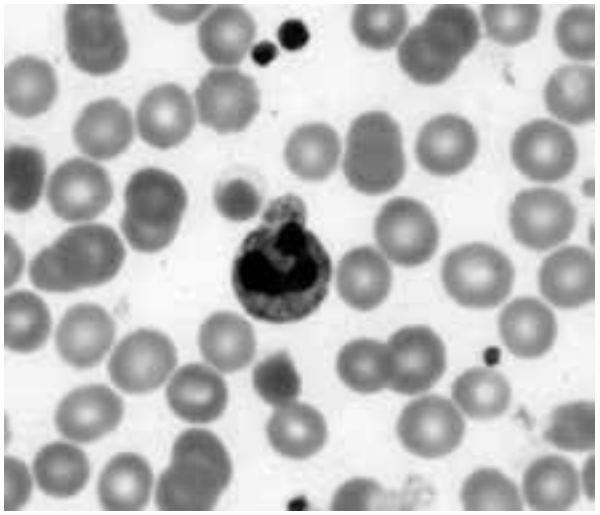
CASE

A 64-year-old women of mixed German and Scandinavian descent presented to the emergency department with a two-day history of abdominal pain, diarrhoea and dark urine. There was no fever present. One month before, she had travelled to Brazil for which no malaria prophylaxis was recommended. Physical examination revealed an icteric women with a body temperature of 36.4°Celsius. Palpation of the upper abdomen was slightly painful. Hepatosplenomegaly was absent.
Laboratory results showed a Coombs-negative haemolytic anaemia (haemoglobin 6.4 mmol/l, reticulocytes 206 x 10^9/l, haptoglobin <0.08 g/l and lactate dehydrogenase 744 U/l) with a thrombocyte count of 203 x 10^9/l and a leukocyte count of 13.4 x 10^9/l. C-reactive protein was 25 mg/l. In *figure 1* the blood smear is shown.

Figure 1. *Blood smear*



WHAT IS YOUR DIAGNOSIS?

See page 487 for the answer to this photo quiz.

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ANSWER TO PHOTO QUIZ (PAGE 483)

A DIAGNOSIS ON THE BASIS OF A BLOOD SMEAR

Prompt blood smear examination showed mainly blister cells. Blister cells are characteristic of acute haemolysis induced by oxidative stress. Because of the oxidative denaturation of the haemoglobin, it is piled at one side of the cell.[1,2] These blister cells raised a strong suspicion on the presence of a glucose-6-phosphate dehydrogenase (G6PD) deficiency. G6PD deficiency was confirmed by decreased erythrocytic G6PD activity (5 U/gram Hb).

This X-linked hereditary disorder mainly affects people from Mediterranean, African, Middle Eastern and South Asian descent.[3] Reduced G6PD concentrations render erythrocytes susceptible to haemolysis under oxidative conditions induced by oxidant drugs, infection, chemicals such as naphthalene or ingestion of fava beans.

Our patient had a urinary tract infection and had eaten fresh fava beans two days before presentation. During the trip to Brazil, she had put mothballs in her pocket. Mothballs with naphthalene are not produced anymore. This case is remarkable for the German/Scandinavian descent and the age of first symptoms. First presentation at an older age can be due to X-chromosome inactivation.[4]

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