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C/β0 thalassemia

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C/β0 thalassemia

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Morphology Update

C/β^0 thalassemia

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The blood film illustrated is from a 60-year-old Afro-Caribbean woman with compound heterozygosity for haemoglobin C and β^0 thalassemia. She had required a cholecystectomy for gall stones at a young age but otherwise had had only minor symptoms. Her spleen was palpable on inspiration. Hemoglobin concentration (Hb) and red cell indices were: red cell count 4.96 x 10^9/l, Hb 7.8 g/dl, hematocrit 0.25 l/l, mean cell volume 62 fl, mean cell hemoglobin 19.1 pg and mean cell hemoglobin concentration 30.9 g/dl. The reticulocyte count was 173 x 10^9/l, bilirubin 56 µmol/l (normal range 3–17) and serum ferritin 431 µmol/l (normal range 15–300).

The blood film shows a haemoglobin C crystal (centre right) and a poorly hemoglobinized erythroblast (centre left). In addition there are target cells and numerous irregularly contracted cells. Pappenheimer bodies were also present. Haemoglobin C crystals are very distinctive because of their straight parallel edges. They are often in an erythrocyte that is otherwise empty of hemoglobin. They can be seen in hemoglobin C disease and in compound heterozygous states when hemoglobin C is co-inhertited with either hemoglobin S or β thalassaemia. The combination in this patient of features of hemoglobin C (crystals, target cells and irregularly contracted cells) with features of thalassemia (microcytosis, poor hemoglobinization and Pappenheimer bodies) points to the correct diagnosis.

For Peer Review