Photoclinic

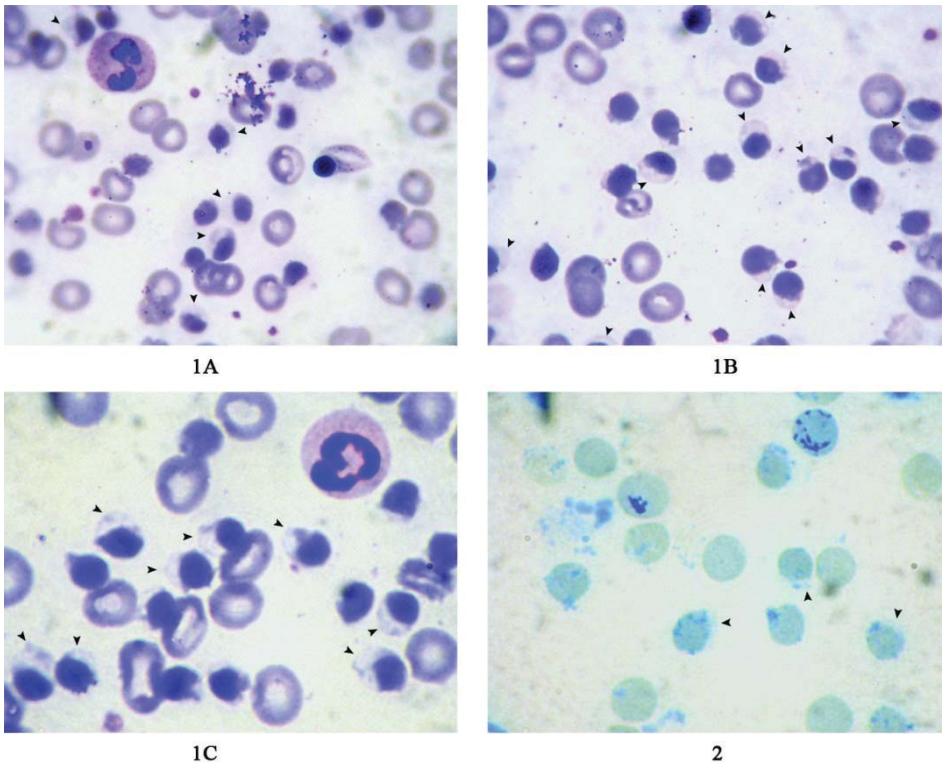

Figure 1 and 2. 1) Peripheral blood smear during hemolytic attack shows many blister cells (hemi-ghost cells) (arrowheads), as well as nucleated RBCs, and spherocytes. (Wright, ×1000). **2)** Supravital staining reveals the presence of Heinz (arrowhead) in addition toreticulocytes (two cells seen in background) (Brilliant cresyl blue stain,×1000).

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A 68-year-old man, a known case of G6PD deficiency, presented to the emergency department with jaundice, flank pain, and "dark urine". On physical examination, the patient was icteric with pale conjunctiva and mild splenomegaly. The examination was otherwise normal and he had not taken any medication. His past medical history was significant for "intermittent jaundice" in the past. The urine analysis was positive for hemoglobinuria and bilirubin pigments with no RBC. The hemoglobin level was 6.0 g/dL (hematocrit: 20.1% with RBC count: 2.06×10^{12}/L) and the reticulocytes count was 6.2%. A peripheral blood smear was obtained (Figures 1 and 2).

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What is your diagnosis?
See the next page for diagnosis.

Photoclinic Diagnosis: **Hemi-ghost Cells**

Hemolytic crisis in G6PD deficiency probably due to ingestion of fava beans.

G6PD deficiency, an X-linked disorder, is the most common disease caused by enzymopathy. This enzyme helps in detoxification of free radicals and peroxides. The most common presentation in adults is anemia caused by hemolytic crisis. It is caused by factors such as oxidative drugs or chemicals, infection, or ingestion of fava beans (favism). Although all patients with favism have G6PD deficiency,[1–2] it is mostly implicated in the Mediterranean region, including the Middle East (where the patient originated).[3–4]

In G6PD deficiency, hemolysis leads to a drop in hemoglobin level and appearance of red cell fragments, blister cells (hemi-ghost cells) (Figure 1A, 1B, 1C arrowheads), anisocytosis, polychromasia, nucleated RBCs, and reticulocytosis. Denatured globin chains often attach to the red cell membrane and can be seen on supravital staining as Heinz bodies inside the blisters protruding into the cytoplasm (Figure 2 arrowheads) with lighter shade of blue than the reticulofilamentous material of reticulocytes. They are consequently removed by splenic macrophages, producing bite cells. On further questioning, the patient recalled eating fava the day before. He was treated conservatively and the follow up period was uneventful.

References

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