Educational Material IQAP 1331

A 57-year-old man was admitted to a hospital for investigation of chest discomfort and treatment of atrial fibrillation and heart failure. A complete blood count demonstrated Hb 8.5 g/dL, RBC 2.68 $x10^{12}$/L, Hct 0.262 L/L, MCV 97.8 fL, WBC 20.8 $x10^{9}$/L and platelet 168 $x10^{9}$/L. His haemoglobin level dropped progressively after admission. He presented hyper-bilirubinaemia, depressed haptoglobin concentration and elevated LDH level. Upon investigation, the patient was noted to be Glucose-6-phosphate dehydrogenase (G6PD) deficiency and have taken herbal medication before admission.

The blood film shows a leuco-erythroblastic blood picture with polychromasia (Figure 1). There are many nucleated red cells and a small number of myelocytes. A marked degree of poikilocytosis with irregularly contracted cells, blister cells, helmet cells and bite cells is noted (Figure 2). Neutrophilia with left shift is also evident. The overall features are compatible with the diagnosis of oxidative haemolysis.

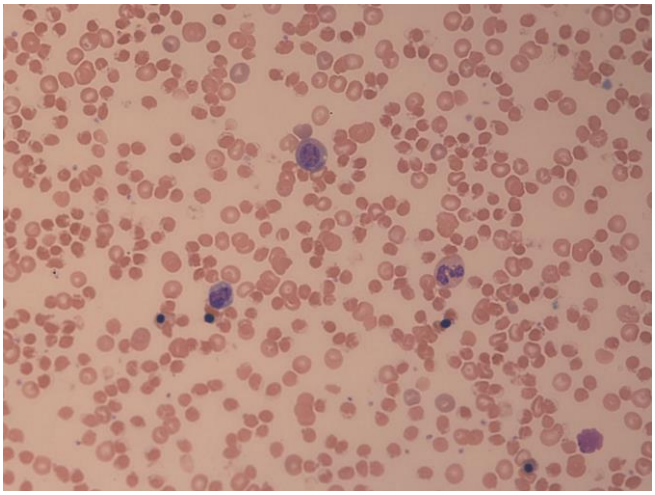

Figure1. Leucoerythroblastosis with polychromasia showing the presence of nucleated red cells and myeloid precursors. (10x magnification)

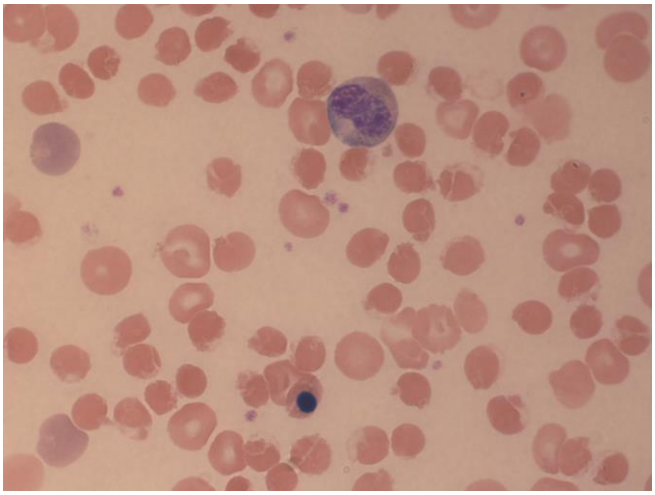

Figure 2. Contracted cells, blister cells, helmet cells and bite cells are suggestive of oxidative stress-mediated haemolytic anaemia. (100x magnification)

G6PD is one of the enzymes enabling erythrocytes to physiologically cope with oxidative stress. There are several common genetic variants of G6PD deficiency namely, $G6PD^{A-}$, $G6PD^{mediterranean}$ and $G6PD^{Canton}$. The production of super-oxides from oxidative stress converts haemoglobin to methaemoglobin. In the absence of G6PD, methaemoglobin interacts with hydrogen peroxide. Heinz bodies are formed and they cling to the surface of red cell membrane, which becomes rigid and subject to sequestration in the spleen, resulting in the formation of helmet cells and bite cells. The characteristics of red cell morphology in the blood film are suggestive of the diagnosis of the disease. Heinz bodies can be demonstrated at time of acute haemolysis, whereas enzymatic assays of G6PD at acute haemolysis may be aberrantly normal, which is attributed to the up-regulated enzymatic activity in some G6PD variants with reticulocytosis in an attempt to compensate the haemolytic anaemia.