

Case of Acquired Stomatocytosis in an Alcoholic Patient

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ABSTRACT

We present a case of acquired stomatocytosis in red blood cells of a middle aged alcoholic male, who presented with mild anemia. Stomatocytes are erythrocytes with a central slit or mouth-shaped area a central pallor. The pathology lies in decreased activity of sodium potassium membrane pump leading to increased sodium influx causing cell swelling inducing stomatocytosis. The increased dependence on automated hematology analyzers and minimum use of peripheral blood smear examination in modern time necessities the need of peripheral blood smear examination.

Keywords: Stomatocytes, alcoholism, peripheral blood examination

Introduction

Stomatocytes are erythrocytes with a central slit or mouth-shaped (stoma) area of central pallor when examined on dried smears. A few stomatocytes may be observed in blood smears prepared from normal individuals[1]. Stomatocytes are associated with aberrant Rh blood-group antigen expression, sitosterolemia, and familial deficiency of high-density lipoproteins.[2]

The major pathophysiologic abnormality is the result of a marked increase in sodium permeability (15 to 40 times normal) leading to increased RBC sodium, a lesser decrease in intracellular potassium, an increase in total monovalent cation content, and, thereby, an increase in cell water [3]. Despite a marked compensatory increase of active sodium and potassium transport, increased pump activity is unable to compensate for the markedly increased inward sodium leak. Red cells swell when the inward sodium leak exceeds the potassium leak out; red cells shrink when the potassium leak out exceeds the inward sodium leak. These abnormalities are recognized by observing changes in MCHC (cell hydration) [4].

Case Report

A 62 year male presented with complaints of sudden onset of breathlessness, progressive abdominal swelling, black stools and swelling in lower limbs since 15 days. There was no history of fever, chest or abdominal pain. The past history revealed jaundice, 8 years back and surgery for inguinal hernia 6 years back. There was history of smoking since 20 years and alcoholism since 15 years (about 90-100 ml of whisky everyday). The rest of family history along with patient past medical history was unremarkable. The general examination showed

pedal edema and enlarged parotid gland. Pallor, Icterus, Cyanosis, clubbing and lymphadenopathy were not seen. Respiratory system showed decreased bilateral air entry with rhonchi. Cardiovascular examination revealed a pansystolic murmur. The other systemic examination was unremarkable. The vitals were; Blood pressure 120/80 mm of Hg, pulse rate 76/minute and temperature – 98.60F. The initial investigations showed hemoglobin level of 11.9 g/dl, mean cell volume (MCV) 121.6 fl, total leucocytes count 11220 / mm³, with neutrophils-76%, lymphocytes-22%, monocytes-2%, platelet count- 1,50,000 / mm³ and erythrocyte sedimentation rate (ESR) 14 mm/ hr. RBC showed central pallor with slit like spaces ? stomatocytes. (Fig-1,2) Reticulocyte count was 1.2% and Prothrombin time was 14.9 seconds (INR-1.13). Biochemical investigations showed Blood urea nitrogen (BUN)- 39 mg/dl, Creatinine 1.2 mg/dl, SGOT- 38 IU, SGPT-31 IU, Total bilirubin- 6.0 mg/dl, direct bilirubin- 0.8 mg/dl, total protein- 6.8 gm/dl, Albumin-3.5 gm/dl, globulin- 3.3 gm/dl, A/G ratio- 1.06, Alkaline phosphatase 73 IU, Gamma GT- 24. The electrolytes were within normal limits, Sodium- 142mEq/L, Potassium- 4.2 mEq/L and chloride 93 mEq/L. The patient tested non reactive for HIV-1 & 2, Hepatitis B surface antigen, hepatitis C antibody , Ig M anti HbS. Urine examination revealed a pale clear appearance with 0-1 / hpf RBC , 5-6/ hpf WBC ,occasional epithelial cells and absence of any cast, crystal or bacteria.

Discussion

Stomatocytes are red cells with central slit like spaces seen in healthy normal individuals and when in large numbers is the cause of varying degree of hemolysis by altered sodium permeability of the red cell membrane [5, 6]. It can be acquired or genetic. Acquired stomatocytosis

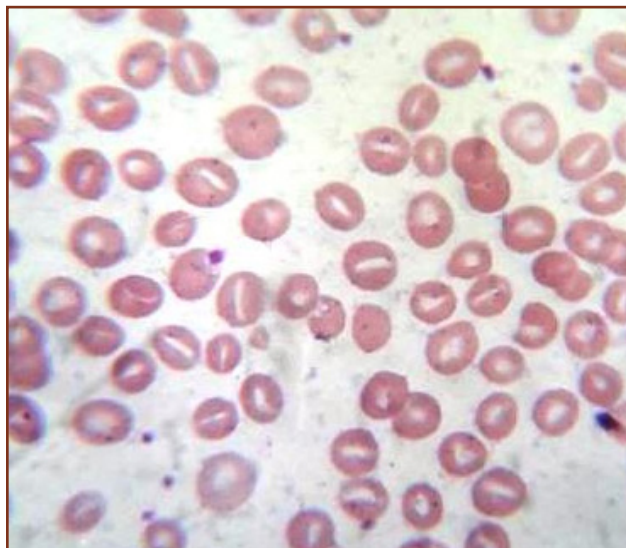


Fig. 1: Slit like spaces in RBC- Stomatocytes (Leishman stain, 100 x oil immersion).

is associated with acute alcoholism and hepatobiliary disease, vinca alkaloid administration, neoplasm, cardiovascular disease and as a processing artifact. The genetic variant has an autosomal dominant mode of inheritance. The excess cation permeability is associated with an absence of red cell membrane protein -band 7 on sodium dodecyl sulfate gels, this protein is referred to as band 7.2 b or stomatin [7,8]. In knockout mice model stomatin deficiency did not result in morphological aberration in form of stomatocytes therefore making the function of the gene uncertain [9, 10]. Cells treated with dimethyl adipimidate normalize membrane permeability and corrects the abnormal RBC morphology.

The severity of hemolytic disease is diverse, both between different families and among affected members of the same family. In most patients, symptoms related to intermittent anemia and jaundice are so mild that no therapy is required. Rarely, the anemia is of sufficient severity to require transfusion therapy. Splenomegaly is an expected corollary of severe anemia. The MCV is elevated, often strikingly, the MCHC is normal or low (as a result of increased cell water content), and osmotic fragility is increased. Red cell sodium content is increased and potassium content is decreased. In most patients, hemolytic anemia is improved after splenectomy, but splenectomy is considered contraindicated in most cases because there is a high risk of post splenectomy thromboembolic disease and chronic pulmonary hypertension. This has been attributed to increased erythrocyte-endothelial cell

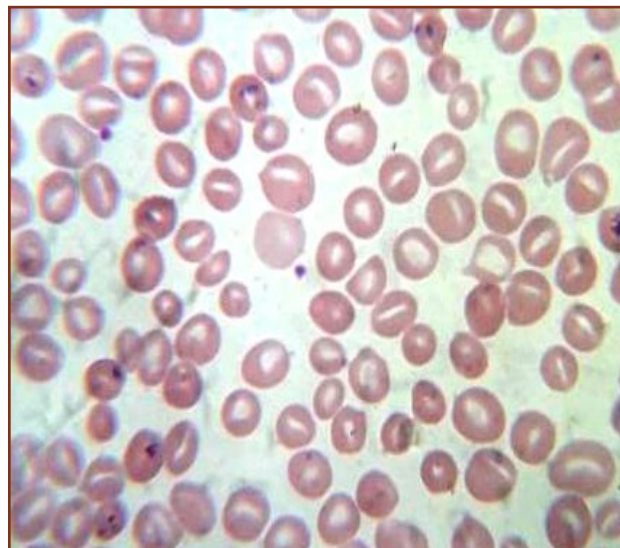


Fig. 2: Stomatocytes (RBC with slit like spaces) seen with normal RBC (Leishman stain, 100 x oil immersion).

adhesion resulting from increased phosphatidylserine exposure of over hydrated erythrocytes fortunately, most patients have compensated hemolysis and splenectomy is not required. [11]

The strong history of alcohol intake was the cause of stomatocytes in peripheral blood smear in our case as there were no other factors responsible for this morphological abnormality seen in our patient.

Conclusion

The trend of modern hematology practice is increased dependence on automated hematology analyzers and minimum use of peripheral blood smear examination. The microscopic examination of slide is the key modality for elucidating such morphological variations and the pathologist must be aware of the importance of reviewing peripheral blood smear.

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