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Unmasking Pseudosickle Cell Anemia: A Case Report of Disseminated Intravascular Coagulation(DIC)-Associated Hematological Anomalies

(Desenmascarando la anemia pseudofalciforme: informe de un caso de anomalías hematológicas asociadas a coagulación intravascular diseminada (CID))

Sai Sudha M [1], J Thanka [1], Evangeline Cynthia Dhinakaran [1], Anbukarasi K [1]

[1] Department of pathology, Sree Balaji Medical college and Hospital, Chennai, India.

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Abstract(english)

Sickle cell changes in red cells are inherited disorders where the normal concave shaped red blood cells are converted to sickle shaped cells due to genetic mutations in the cell membrane. Pseudo sickle cell shaped red blood cells in a peripheral smear are deformed red cells which mimic the sickle cells but lacks the genetic abnormality.These pseudo sickle cells should be viewed with many differential diagnosis. we report a 75/female with a history of traumatic intertrochantric fracture of the femur, her peripheral smear showed an incidental finding of pseudosickle cells in the peripheral smear and positive sickling test. Hemoglobin electrophoresis was done, it revealed normal hemoglobin. .A detailed hematological work up was done and clinically patient was diagnosed with DIC. Pseudosickling in the backdrop of DIC is a rare entity which required further studies.

Keywords(english)

Pseudo sickle cell, Hemoglobin electrophoresis, Disseminated intravascular coagulation, Sickling test.

Resumen(español)

Las alteraciones de células falciformes en los glóbulos rojos son trastornos hereditarios en los que los glóbulos rojos normalmente cóncavos se transforman en células falciformes debido a mutaciones genéticas en la membrana celular. Los glóbulos rojos con forma de pseudocélulas falciformes en un frotis periférico son glóbulos rojos deformados que imitan a las células falciformes pero carecen de la anomalía genética. Estas pseudocélulas falciformes deben considerarse con muchos diagnósticos diferenciales. Presentamos el caso de una mujer de 75 años con antecedentes de fractura intertrocantérica traumática del fémur. Su frotis periférico mostró un hallazgo incidental de pseudocélulas falciformes y una prueba de falciformación positiva. Se realizó una electroforesis de hemoglobina, que reveló hemoglobina normal. Se realizó un estudio hematológico detallado y clínicamente se diagnosticó a la paciente con CID. La pseudofalciformación en el contexto de la CID es una entidad rara que requiere estudios adicionales.

Palabras clave(español)

Pseudoanemia falciforme, electroforesis de hemoglobina, coagulación intravascular diseminada, prueba de falciformación.

✉ **Autor de correspondencia:** Dr. Sai Sudha. M. Department of pathology, Sree Balaji Medical college and Hospital, Chennai, India Email: radha.5054@gmail.com .

Introduction

Sickle cells are crescent or sickle shaped red blood cells seen usually in Sickle cell disease. Pseudo sickle cells can be seen in some rare conditions that distort the shape of the red blood cells[1]. Unlike true sickle cell anemia, which is characterized by the presence of abnormal hemoglobin S(HbS), pseudosickle cell conditions do not involve the specific mutation and reveals normal chromatogram in Hb Electrophoresis and HPLC.

Case report

A 75 year female came with complaints of pain and inability to walk after the history of fall at her residence. Patient diagnosed with left intertrochanteric fracture and planned for elective Bipolar hemiarthroplasty of left hip . Ultrasound abdomen reveals mild splenomegaly. Her hematological parameters revealed Hemoglobin – 9.5g/dl with hematocrit – 27.0%, Mean corpuscular volume – 69.1 fl, Mean corpuscular hemoglobin – 24.2 pg , Mean corpuscular hemoglobin concentration-35.2% , Red cell distribution width -43.3fl , Total leucocyte count -11000 cells/uL and platelet count 30,000 / uL- . Peripheral smear shows sickle shaped RBCs, schistocytes and nucleated RBCs along with Thrombocytopenia. Sickling test with 2% sodium metabisulphite showed increased sickling in the RBCs immediately and also after first hour incubation at room temperature. We have suspected sickle cell trait with thrombocytopenia initially and suggested HB electrophoresis for further confirmation. However, Hb electrophoresis reveals no abnormal hemoglobin. Further blood samples also shows Sickle shaped cells, increased number of schistocytes in the peripheral smear.

We have investigated further to rule out other causes for Pseudo sickle cell anemia, in which Serum iron and Ferritin were normal. Total bilirubin levels are raised -2.5mg/dl. Serum LDH levels(308U/L) are elevated and Coombs test is negative. FDP levels and D-dimer levels(2610ng/dl) are markedly elevated and platelet count is reduced to 20,000/cumm. Finally patient was diagnosed with Disseminated Intravascular Coagulation(DIC) with pseudo sickle cells in Peripheral blood smear, and started supportive treatment with RDP and FFP transfusions.

Discussion

Sickle cell disease is an inherited disorder in which point mutations of globin chain causes abnormal hemoglobin (HbS) . when the red cells containing this abnormal hemoglobin exposed to hypoxic conditions, the biconcave red cells acquire sickle shape[1,2].

Sickle cells are narrow, elongated cells with pointed ends and also crescent shaped. The sickle cells are seen mainly in sickle cell disease, but few conditions like Iron deficiency anemia, Hereditary elliptocytosis, High altitude and Malarial infections also shows sickle shaped RBCs rarely called Pseudo sickle cells. These are sickle shaped RBCs in the peripheral smear with positive

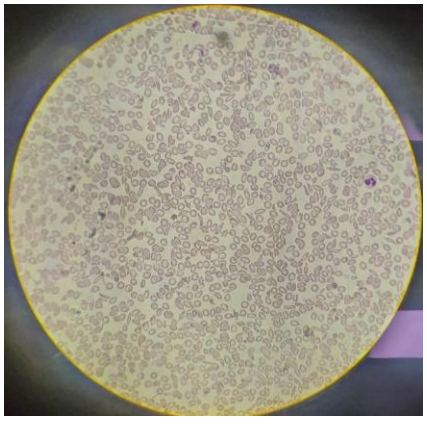

Figure 1. Peripheral blood smear demonstrating numerous sickle-shaped erythrocytes(Blue arrows) and shistocytes (Red arrows) scattered throughout the field, indicating combined features of sickling. Leishman stain, 100x).

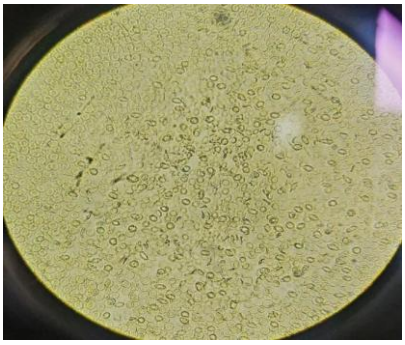

Figure 2. Positive sickling test showing crescent-shaped red blood cells after deoxygenation with 2% sodium metabisulphate. The elongated, curved morphology results from polymerization of deoxygenated HbS, leading to the distortion of normal biconcave RBC shape.

sickling test but the hemoglobin electrophoresis shows normal hemoglobin[3,4]. Rare abnormal Hemoglobins like HbC (Harlem), HbC (Georgetown), HbH, HbI, Hb Barts and Hb Setif also cause Pseudosickling with 2% Sodium metabisulphite[5,6].

Disseminated Intravascular Coagulation causes widespread activation of coagulation factors leads to the formation of fibrin in the microvasculature[7]. This can cause mechanical damage to the Red Blood cells resulting in abnormal shaped RBCs, most commonly Fragmented cells or Schistocytes. Sometimes marked cell distortion causes abnormal shaped RBCs, which might get confused with the sickle cells resulting in Pseudo sickle cell anemia. Pseudo sickling in these cases can be increased by increasing cell distortion and dehydration.

Kasmani et al.[8] and A. Nangia et al[3]. have reported few cases of pseudo sickle cell anemia with Iron deficiency anemia, two cases in elderly female patients and one in case of a male child. Elliptocytes and poikilocytes in the peripheral smear mimic's sickle cells in those cases and later pseudo sickle anemia is confirmed with HB electrophoresis.

This rare case of pseudosickle cell anemia in association with Disseminated Intravascular Coagulation emphasizes the importance of considering secondary causes like DIC, in patients presenting with sickle shaped cells in the absence of sickle cell disease. The accurate differentiation between true sickle cells and pseudosickle cellsis crucial, as the treatment strategies differ significantly.

In conclusion, this case report underscores the need for comprehensive diagnostic workup in case of pseudo sickle cell anemia and highlights the importance of the alternative diagnosis when clinical and laboratory findings do not align with the typical profile of sickle cell disease. Further research is warranted for better understanding the mechanisms leading to pseudo sickle cell anemia in DIC.

Conflict of interest

None to declare.

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