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Case report

Bizarre filamentous of erythrocytes presentation in children with sickle cell disease: about a case

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Abstract

It is a Congolese male child living with homozygous sickle cell disease who has a false thrombocytosis at 988 000/µL and a regenerative hypochromic normocytic anemia with automated hemogram. Peripheral blood smear examination revealed spherical, sperm-shaped or even rosary-shaped red blood cells.

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Introduction

Sickle cell disease is a form of hemoglobinopathy in which there are also normal forms of red blood cells, some of which are found in many other forms called sickle cells. This deformation is most often due to the polymerization of hemoglobin S (HbS) molecules in fibers form in hypoxia [1]. Associated or not to other hemoglobinopathy, the red cells of the sickle cell patient may have other morphological abnormalities; especially target erythrocytes when associated to the Hb C, the microcytes when combined with thalassemia [2,3]. The presentation of this child's red blood cells may suggest the hypothesis of the association of an anomalous membrane or another pathology with hemoglobinopathy.

Patient and observation

We have fortunately in a complete blood count, found in a patient with a major SCD syndrome particular shapes of erythrocytes. This is the blood sample of a 7 year old child living with homozygous sickle cell disease (SCD) we received in the medical laboratory of the National Sickle Cell Disease Reference Center (CNRDR) Brazzaville. The sample taken from tube containing tetraacetic ethylene diamine (EDTA) was stirred for 5 minutes and thereafter hemogram was performed with a cell hematology PLC (AHC) Sysmex XN 350 in CBC + DIFF + RET mode. The results obtained are shown in Table 1. When reading this hemogram, several alarms were signal by the controller. The alarms and other abnormalities observed on the rendering of the AHC led us to perform a blood smear in order to allow us to validate the blood count. The following findings were made: false lymphocytosis was cancelled on the thin blood film after erythroblasts count at 80% of all nucleated cells. This led us to correct the number of lymphocytes and consequently the leukocytes at 7470/µL, excluding therefore leukocytosis; thombocytosis of 988,000/µL was not observed on the thin blood film. It was probably fragmented erythrocytes observed on thin blood film which were falsely counted by the AHC as platelets. The presence of erythroblasts, anisocytosis and fragments of red blood cells was well documented on the smear. Nevertheless the thin blood film enable us to visualize: the presence of red blood cells with sickle-shaped (sickle cell), of schistocytes, cellular debris with various forms (in filament, in pearl, in dot) (Figure 1). These particular presentation of red blood cells in filament form, or pearl, or dot seems unusual.

Discussion

Leukocytosis found in malignant process or not, is also seen during infections of various causes. In sickle cell patients, moderate leukocytosis is very common in both critical and non-critical phases [4,5]. In the latter, because of their chronic anemia, erythropoiesis is very active; which shows in SCD patients a high number of reticulocytes and erythroblasts in the peripheral blood [6]. These erythroblasts, which are nucleated cells, are falsely counted as lymphocytes by most AHCs. Indeed, the non-lobulation of the nucleus of these erythroblasts and the absence of granulation in the cytoplasm cause the AHC to classify them in the group of lymphocytes. Hence leukocytosis due to the false lymphocytosis observed in our case. This underscores the importance of paying attention to the interpretation of AHC alarms and reading the blood smear in order to eliminate false leukocytosis and lymphocytosis [7]. The thrombocytosis observed in SCD patients is due to the inflammatory process in these patients [8]. This same observation was made by Lombardo *et al.* (2017) in the United States of America [9]. A careful reading in our daily practice of blood smears is of great importance in case of all thrombocytosis in patients with sickle cell disease in order to exclude false thromrombocytosis. Sickle cells which

are deformed red blood cells in sickle-shaped following the polymerization of hemoglobin S molecules in hypoxia situation in fibers characterized SCD patients. Associated with other hemoglobinopathies, or other pathologies, sickle cell patient's erythrocytes may have other abnormalities including target erythrocytes when associated with the Hemoglobin C and the microcytes when combined with thalassemia [2,3]. Several authors described in the literature some particular forms of erythrocytes in SCD patients. These special presentations are associated with a worsening of the crisis or other pathology associated to SCD [9,10].

Conclusion

Although thrombocytosis is an integral part of the pathophysiological processes in a sickle cell patient, it is advisable to perform a blood smear in order to confirm this thrombocytosis in case of any thrombocytosis from a cellular hematology automaton.

Competing interests

The authors declare no competing interest.

Authors' contributions

A substantial contribution to data acquisition, analysis and data interpretation have been made by: JLS. The writing of the article and the critical review of its important intellectual content were made by: IK, TLOG, FM, OFGA, SLTT, LGO. The final approbation version to publish was given by: EDA. All the authors have read and agreed to the final manuscript.

Table and figure

Table 1: hemogram

Figure 1: different presentations of X100 blood smears

References

1. Renaudier P. Physiopathologie de la drépanocytose. Transfus Clin Biol. 2014;21(4-5):178-181. **PubMed** | **Google Scholar**

2. Wong P. A hypothesis of target cell formation in sickle cell disease. Med Hypotheses. 2016;93:102-105. **PubMed** | **Google Scholar**

3. Fenneteau O, Hurtaud-Roux M-F, Schlegel N. Aspect cytologique normal et pathologique du sang chez le nouveau-né et le jeune enfant. 2006;64(1):17-36. **PubMed** | **Google Scholar**

4. Thiam L, Dramé A, Coly IZ, Diouf FN, Seck N, Boiro D *et al*. Epidemiological, clinical and hematological profiles of homozygous sickle cell disease during the intercritical period among children in Ziguinchor, Senegal. Pan Afr Med J. 2017;28:208. **PubMed** | **Google Scholar**

5. Curtis SA, Danda N, Etzion Z, Cohen HW, Billett HH. Elevated steady state WBC and platelet counts are associated with frequent emergency room use in adults with sickle cell anemia. PLoS One. 2015;10(8):e0133116. **PubMed** | **Google Scholar**

6. Oliner HL, Heller P. Megaloblastic erythropoiesis and acquired hemolysis in sickle-cell anemia. N Engl J Med. 1959;261(1):19-22. **PubMed** | **Google Scholar**

7. Geneviève F1, Godon A, Marteau-Tessier A, Zandecki M. Automated hematology analysers and spurious counts Part 2, Leukocyte count and differential. Synthèse Ann Biol Clin. 2012;70(2):141-54. **PubMed** | **Google Scholar**

8. Zhang D, Xu C, Manwani D, Frenette PS. Neutrophils, platelets and inflammatory pathways at the nexus of sickle cell disease pathophysiology. Blood. 2016;127(7):801-809. **PubMed**

9. Lombardo JL, Lynch DT. Sickle cell trait with β-thalassemia, elliptocytosis and thrombocytosis. Blood. 2017;130(10):1275-1275. **PubMed**

10. Siow W, Matthey F, Bain BJ. The significance of irregularly contracted cells and hemighosts in sickle cell disease. Am J Hematol. 2017;92(9):966-967. **PubMed** | **Google Scholar**

Table 1: hemogram

Parameter	**Value**	**Unit**
Leucocytes	37. 3 x 103	µL
Erythrocytes	2.54 x 106	µL
Hemoglobin	6.1	g/dL
Hematocrit	20.9	%
Mean corpuscular volume	82.3	fL
Platelets	988 x 103	µL

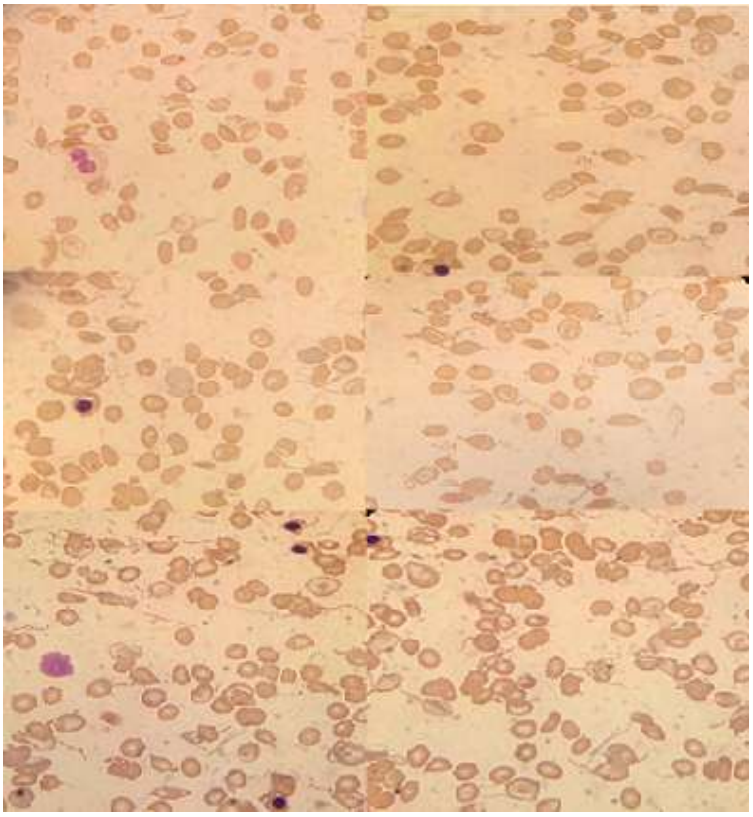

Figure 1: different presentations of X100 blood smears