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Case Report

Urinary Sickle Cells in a Patient with Vaso-Occlusive Crisis: A Case Report

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Abstract

Vaso-occlusive crisis is the most common complication in sickle cell disease and can cause multi organ damage. Here we present a case of sickle cell anemia with vaso- occlusive crisis. The patient is a 39-year-old tribal male, a known case of homozygous sickle cell disease who presented with acute respiratory infection, bilateral lower limb pain and decreased urine output. Peripheral blood smear showed sickled red cells with features of hemolysis and reticulocytosis. Hemoglobin electrophoresis showed prominent HbS band. His symptoms progressed rapidly and he developed hematuria. Urine examination showed sickled erythrocytes. Later the patient went into cardiac arrest and succumbed to the disease. While hematuria is a common symptom in sickle cell disease, sickled RBCs in urine microscopy should be considered a bad prognostic indicator.

Keywords: Sickle Cell Anemia; Hematuria; Urine Microscopy; Sickled Erythrocytes

Abbreviations

SCA: Sickle Cell Anemia; SCD: Sickle Cell Disease; HPLC: High Performance Liquid Chromatography; IV: Intravenous

Case Presentation

A 39-year-old tribal male patient was admitted in a tertiary care center in Kerala with complaints of fever, cough, dyspnea, abdominal pain and bilateral lower limb pain. He had difficulty in walking with history of recurrent falls. He also complained of decreased urine output for the past two to three days. He was a known case of type 2 diabetes mellitus and coronary artery disease and a past history of pulmonary tuberculosis, treated with anti-tubercular drugs. He was diagnosed with SCA and was on treatment since 5 years. However, he had stopped medications for the past 2 months. High Performance Liquid Chromatography (HPLC) done 5 years back had showed 79.5% HbS, 17% HbF, 3.5% HbA2, and no HbA, suggestive of HbS (sickle cell homozygous) disease. He also had a similar history of pain over both legs 6 months back that resolved with treatment. On admission patient was tachypneic with visible pallor and icteric sclera. Chest auscultation showed bilateral crepitations. Abdominal examination showed guarding and rigidity. Yellowish discoloration of urine was also noted. Routine blood investigations showed mild anemia, along with leukocytosis and thrombocytosis. Elevated urea (293 mg/dL), creatinine (3.6 gm/dL), hyponatremia and hyperkalaemia (7.3 meq/L) were also noted. Liver function test showed hyperbilirubinemia, grossly elevated transaminases and prolonged PT and APTT levels. Ultrasound abdomen was unremarkable with normal sized liver and spleen. Peripheral smear showed sickled red cells with hemolysis, anisopoikilocytosis and neutrophilic leukocytosis suggestive of sickle cell crisis (Figure 1). Reticulocyte count was 10%. Hemoglobin electrophoresis showed prominent HbS band (Figure 2). Treatment was started with Intravenous fluids (IV), analgesics and IV antibiotics. Fresh frozen plasma was also transfused.

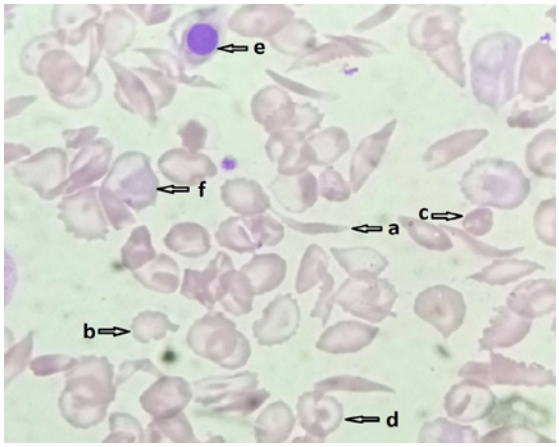

Figure 1: Peripheral smear showing RBC anisopoikilocytosis with numerous **a)** sickle cells, **b)** microcytes, **c)** microspherocytes, **d)** target cells, **e)** nucleated RBCs, **f)** polychromatophilic cells.

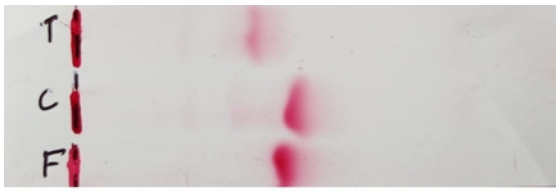

Figure 2: Hemoglobin electrophoresis showing prominent HbS band. Test (T)-HbS, Control (C)-HbA and Fetal blood (F)-HbF.

On the next day, patient developed hematuria and his symptoms worsened. Urine microscopy showed numerous sickled erythrocytes (Figure 3). He was planned for hemodialysis in view of hyperkalemia, rising creatinine levels and worsening dyspnea. Unfortunately, patient had sudden saturation drop along with cardiac arrest and

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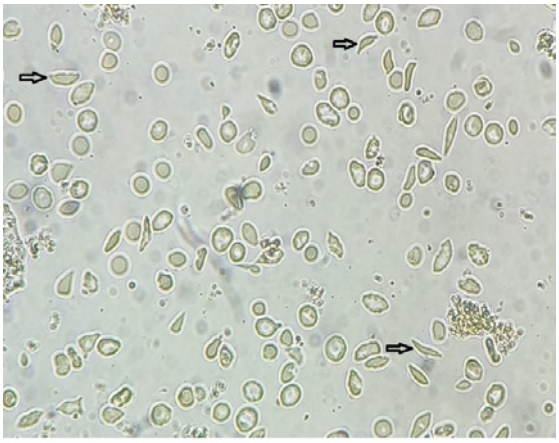

Figure 3: Urine microscopy showing numerous sickled erythrocytes (arrows).

succumbed on the same day.

Discussion

Sickle cell anemia is an autosomal recessive disorder developed as a result of missense mutation at the 6th position of beta-globin chain where glutamic acid is substituted by valine [1]. The sickle cell morphology is rendered by the polymerization of the deoxygenated HbS into long strands [2]. Risk factors of sickling include hypoxia, dehydration, acidosis, infections and high osmolarity [3]. The most important complication of Sickle Cell Disease (SCD) is acute painful crises, which include vaso-occlusive crisis, acute chest syndrome, aplastic crisis, hyper-hemolytic crisis and splenic sequestration crisis [3]. Among these, vaso-occlusive crisis is the commonest presentation and a leading cause of death [4]. In the capillaries, when the erythrocytes release oxygen, they deform and get trapped in microcirculation. This is the pathophysiology behind acute painful crises [3]. SCD can also manifest as acute multi organ failure, which is a life-threatening complication.

The highest incidence of SCD is reported in equatorial Africa with 40-50% members, in the eastern part of the continent. It has also been reported in United States, South Turkey, Saudi Arabia, Israeli Arabs, and North Mediterranean shore [5]. In India, the geographical distribution of SCD is in central and western regions [6] and is more prevalent in the tribal community. Approximately 8.6% of the total population of India is tribal community [2] and the prevalence of sickle cell gene in them is 5-34% [6]. SCD was first described in the tribes of Nilgiri hills in South India in 1952 [2]. In a study conducted in an ethnic population called Irula in Southern India, sickle hemoglobin was found among 31% of the members [7]. The prevalence of HbS gene in these tribal population in the state of Kerala is also very high (18.2-34.1%) [2]. Till now, only a few cases of sickle cell crisis accounting less than 20 have been reported in India.

This patient is an inhabitant of Nilgiris of South India and belongs to Irula community. His clinical presentation along with laboratory findings favor the diagnosis of vaso-occlusive crisis. The risk factors could be the cessation of medication and possible lower respiratory infection. In the terminal stage, patient also had hematuria with presence of sickle cells in urine microscopy. Hematuria is a common symptom in SCD. But the presence of sickled erythrocytes in urine is a very rare finding. Sickle cell nephropathy is also a major complication of sickle cell disease. Sickle cell nephropathy is a group of renal complications consisting of renal papillary necrosis, hematuria, hyposthenuria, acute and chronic kidney injury, proteinuria, sickle cell glomerulopathy and renal tubular disorders [8]. The renal medulla provides all the favourable conditions for RBC sickling due to its high osmolarity, decreased oxygen tension and acidosis. The exact mechanism of renal papillary necrosis is not known, [9] but the vaso-occlusive phenomena and the resultant medullary ischemia might have contributed to it. The evidence of sickled erythrocytes in urine shows rapidly progressive renal failure and hypoxemia which ultimately culminated in the death of the patient.

Conclusion

The appearance of sickle cells in the urine of SCD patients must be considered as a bad prognostic factor. Any patient presenting with hematuria, which is a common symptom in SCD, should be investigated for sickle cells in urine microscopy and managed aggressively.

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