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Symmetric Peripheral Gangrene (SPG) of Four Limbs in a Child with Sickle Cell Anemia

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Case Report

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Abstract: Sickle cell disease (SCD) is the most common genetic disorder in the world, which has a varied clinical manifestations and severity. Vascular thrombosis leading to symmetrical peripheral gangrene is rare complication in patients with SCD. We report a 2year old boy with sickle cell anemia who developed symmetrical peripheral gangrene of four limbs following pneumococcal sepsis. Child was successfully managed with antibiotic and supportive care. The gangrenous part of extremities separated and healing follows gradually.
Keywords: Symmetrical, Peripheral gangrene, Sickle cell anemia, Children.

INTRODUCTION

Symmetric peripheral gangrene (SPG) is a well-known but rare clinical syndrome characterized by distal ischemic injury leading to gangrene of two or more sites in the absence of large vessel obstruction [1]. This can be caused by infectious and non-infectious causes with DIC almost always at the background [1, 2]. Sickle cell disease has been recognized as hyper-coagulable states and patients with SCD exhibit high level markers of thrombin generation molecules such as D-Dimers, PAP complexes and fibrinopeptide A [3-5]. In addition, there is a decreased level of natural anticoagulant proteins (protein C and S) [6], Platelets are both chronically elevated and activated [7, 8]. These manifest clinical thrombotic complications like pulmonary embolism, stroke and peripheral gangrene [7].

We report a case of a 2year old boy known sickle cell anemia, who presented with SPG of the four limbs complicating streptococcal sepsis.

CASE REPORT

A 2year old boy with SCA presented with fever, pain and dark discoloration of both feet and hands, extending to the forearm. He had vomiting and diarrhea for 5 days, which stopped 2days before presentation. The child is diagnosed to have SCA at 6 months of age with no prior history of hospitalization or blood transfusion. The patient has not received hemophilus influenza type b and pneumococcal conjugate vaccines. At the initial onset of his the illness, he received medications from a pharmacy store including oral amoxicillin, and acetaminophen.

On examination child was irritable, pale, febrile (38.9^0C axillary temperature), no bleeding from any orifice no petechial or purpuric rashes. There were symmetrical black discolorations of all digits of hands and feet, with mixed areas of hyperemia, hyper pigmentation and bullous eruptions involving the palms and dorsi of the hands and feet extending up to the proximal one third of the fore-arms and legs(Figure 1). Peripheral pulses were palpable. Other systems were stable. His CBC on admission showed Hb 5.4g/dl, WBC 54,000/mm^3 and platelet count 1,034,000/mm^3. Kidney and liver function tests were normal. Upper and lower limb arterial Doppler scan showed small vessels vasculitis. Blood culture revealed Streptococcus pneumonia. The child received packed red cell transfusion, Fresh frozen plasma, subcutaneous low molecular weight heparin (clexane), intravenous antibiotics (ceftriazone), and intravenous fluid. Orthopedic surgical team was consulted who suggested disarticulation when the progression has stopped. On the 5^{th} day of admission, fever subsided and repeat CBC showed haemoglobin (Hb) 8.7g/dl, WBC 20,000/mm^3 and platelet count dropped to 154,000/mm^3. Progression of the gangrenous process also stopped. The parents refused amputation. The discoloration of the finger and toes receded, followed by auto-amputation of the digits and patient was subsequently discharged and he is currently stable on follow up.

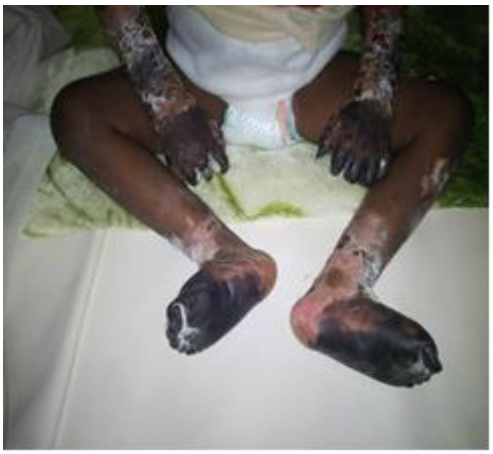

Fig-1: Symmetric peripheral gangrene of the four limbs with purpura fulminans in a child with SCA

DISCUSSION

Although symmetrical peripheral gangrene (SPG) in patients with SCD is well-documented in the literature, it appears to be a rare complication [2, 9]. SPG can be caused by infectious and non-infectious causes with DIC almost always at the background. Individuals with SCD have increased susceptibility to invasive bacterial infections which has been attributed to variety of reasons including dysfunctional antibody production and opsonophagocytosis, hypo-complimentaemia and defective splenic clearance [10]. Newborn screening for early diagnosis and entry of diagnosed children into SCD comprehensive care and treatment programs, including immunization against invasive pneumococcal disease and oral penicillin prophylaxis, coupled with education about detecting splenic sequestration and early warning signs of infection will alleviate morbidity from bacteremia in children aged less than 5 years. Unfortunately, in sub-Saharan Africa where majority of children affected by SCD live, newborn screening and comprehensive care program for children under 5 years of age has not been widely implemented. Hypercoagulation has also been described as prominent feature of SCD and is mediated by activation of both intrinsic and extrinsic coagulation pathways, and this may contribute to thrombotic complications [8, 11]. .Other factors that contribute to developing gangrene in SCD include tendency to form clots from deformed red cells, injured vessels, and decreased nitric oxide bioavailability leading to micro-vascular occlusion [9]. Gangrene in patients with SCD once established rarely managed by embolectomy or revascularization, leaving amputation as the only alternative [10, 11]. Our patient developed gangrene of the four extremities which is rare and devastating clinical presentation complicated pneumococcal sepsis. Home remedies of vaso-occlusive crises or dactylitis like dipping the limb in cold water is a known precipitating factor of gangrene [12]. The parents of our patient denied the use of such intervention. Immunization against invasive bacterial infection with pneumococcal conjugate vaccine and oral penicillin prophylaxis couple with early recognition and prompt treatment of infection in children with SCD will significantly reduce morbidity and mortality.

REFERENCES

1. Kumar P, Tansir G, Gupta H, Dixit J. Symmetrical peripheral gangrene in severe Plasmodium vivax malaria. Indian journal of critical care medicine: peer-reviewed, official publication of Indian Society of Critical Care Medicine. 2017 Apr;21(4):245.
2. Ghosh SK, Bandyopadhyay D. Symmetrical peripheral gangrene. Indian J Dermatol Venereol Leprol 2011;77:244-8
3. Sparkenbaugh E, Pawlinski R. Interplay between coagulation and vascular inflammation in sickle cell disease. Br J Haematol. 2013 Jul; 162(1):3-14.
4. Ataga KI, Key NS. Hypercoagulability in sickle cell disease: new approaches to an old problem. Hematology Am Soc Hematol Educ Program. 2007;91-6.
5. Setty BN1, Rao AK, Stuart MJ. Thrombophilia in sickle cell disease: the red cell connection. Blood. 2001 Dec 1; 98(12):3228-33.
6. Schnog JB, Mac Gillavry MR, van Zanten AP, Meijers JC, Rojer RA, Duits AJ, ten Cate H, Brandjes DP. Protein C and S and inflammation in sickle cell disease. Am J Hematol. 2004 May; 76(1):26-32.
7. Ibijola AA, Ojo OT, Shokunbi WA. Evaluation of Protein C in Nigerian Patients with Sickle Cell Anaemia in Steady State. BJMMR, 2016; 11(9): 1-5.
8. Gladwin MT, Kato GJ. Hemolysis-associated hypercoagulability in sickle cell disease: the plot (and blood) thickens! Haematologica. 2008 January; 93(1): 1–3.
9. Kurhade A K, Lakhkar B B. Peripheral Gangrene in Sickle Cell Disease- A Rare but Dreaded

Complication. Pediatric On-call [serial online] 2016[cited 2016 October-December.
10. Battersby AJ, Knox-Macaulay HHM, Carrol ED. Susceptibility to Invasive Bacterial Infections in Children with Sickle Cell Disease. Pediatr Blood Cancer 2010; 55:401-406.
11. Michaels LA, Maraventano MF, Drachtman RA. Thrombosis and gangrene in a patient with sickle cell disease and dactylitis. J Pediatr 2003; 142:449.
12. Saleem S, Rice L. Limb Amputation in Hemoglobin SC Disease after Application of Ice and Elevation. Am J Hem 2007; 82:53–54.