

**Case Report****Sickle Cell Nephropathy Presenting as Type 4 Renal Tubular Acidosis and Hyperkalemia**

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DMIMS Univ., JN Medical College, Sawangi (M), Wardha- 442004 Maharashtra**Abstract**

Sickle cell nephropathy (SCN) is a well known clinical entity. Various pathophysiological mechanisms and risk factors alter the renal functions in sickle cell disease (SCD). The renal manifestations ranges from glomerular insults, vasculopathy, tubular dysfunctions and even malignancies like renal medullary carcinoma. We present a case of a 20 year male who presented to us with sickle cell crisis and severe hyperkalemia due to rare type 4 distal renal tubular acidosis (RTA).

Key Words: SCN, SCD, vasculopathy, glomerular, hyperkalemia, RTA.

Introduction

Renal failure in SCD ranges from 5 – 18 % of the total population of SCD patients. ^[1] The median age from the diagnosis of disease to renal failure approximates to 20– 23 years. Proteinuria, hypertension, severe anaemia, hematuria ,metabolic acidosis and electrolyte imbalances are known poor prognostic indicators. ⁽²⁾ Such diverse renal manifestations are due to the complex vascular alterations and the tendency of the red blood cells to sickle in the hypoxic, acidotic and hyperosmolar renal medulla. Distal nephron dysfunction is well known in SCN. It can manifest as hypokalemic, hyperchloremic, non anion gap metabolic acidosis. Rarely, type 4 renal tubular acidosis with hyperkalemia complicates SCN. ⁽³⁾

Case Report

A 20 year old male, a known case of sickle cell disease since 10 years presented to us with fever,

cough and breathlessness of 5 days duration. There was no history of haemoptysis, hematuria, edema, syncope, oliguria.

On general examination, patient was restless, tachypnoeic with deep respirations .Pulse was 122/min, regular. Blood Pressure was 92 /50mm Hg, respiratory rate was 28 cycles /min with accessory muscle of respiration in action. Patient was febrile, Jugular venous pressure (JVP) normal, pallor icterus were present. Respiratory system revealed bronchial breath sounds and crepitation in right inter and infra scapular areas. Investigations revealed – Hb-5.2 gm%, TLC-22,000cells/mm³, with 88% neutrophils. Platelet count was normal, serum bilirubin 5.8 mg/dl with predominant unconjugated fraction. Serum LDH-2500 U/L.

Chest X ray revealed consolidation in right lower zone. ABG – PH=7.108, PaCo2=30 mm Hg, PO2= 90 mm Hg , HCO3 = 8mEq/L, SO2 = 95 %

suggestive of severe metabolic acidosis with normal anion gap. Serum potassium was 6.8 mEq/L. Serum urea- 38mg/dL. serum creatinine was 1.9mg/dL

The patient was treated with intravenous antibiotics, 2 units of blood transfusion, hydration, injection soda bicarbonate, inj calcium gluconate, IV inj dextrose insulin drip but the metabolic acidosis persisted with hyperkalemia for which patient was taken for hemodialysis. After two sessions of dialysis potassium level came back to normal, patient's condition improved over next 5 to 6 days. The repeat chest X ray showed clearing of consolidation. The patient was discharged on hydroxyurea 1gm, oral iron, folic acid and zinc supplementation.

Discussion

Kidney involvement in SCD presents with myriad of glomerular and tubular disorders increasing the chances of morbidity and mortality^(4,5) The pathophysiology being polymerization of erythrocytes into renal medulla, a region, predisposing to this crisis because of low local oxygen pressure, low pH and high osmolality.^(5,6) These alterations lead to tubular functional abnormality.

Impaired medullary perfusion causes distal nephron dysfunction, this manifests as acidification defect with or without impaired ability to secrete potassium.

Evidence suggests that in a steady state SCD distal renal tubular acidosis coexists with mild respiratory acidosis in some patients.^(7,8) Medullary ischaemia in SCN impairs the energy dependent process that maintains an adequate proton gradient for acid excretion. A study on 400 patients of SCD showed that 40% had RTA which was normokalemic, possibly due to diminished availability of ammonia.⁽⁹⁾

Impaired potassium excretion in SCN can rarely occur due to resistance of distal nephron to aldosterone.⁽¹⁰⁾ Medullary ischaemia can also contribute to the same. Very rarely like in our case hyperkalemic hyperchloremic metabolic acidosis (type 4 RTA) occurs in SCD. Hyporenemic

hypoaldosteronism contributes to the same in some cases.⁽¹¹⁾

Conclusion

The present case demonstrates the clinical significance of a rare variety of RTA presenting as hyperkalemia in SCD patients. While treating metabolic acidosis in SCD, serum potassium plays a very important role for distinguishing type 4 RTA from other commonly occurring renal tubular disorders. This should be kept in mind while treating SCN.

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