Sickle Cell Disease with Left Elbow Monoarthritis: A Typical Presentation

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Abstract: Sickle cell disease (SCD) is worldwide distributed most common type of inherited blood disorder characterized by the vaso-occlusive crises due to change in the shape of RBC successive to the polymerization of one type of hemoglobin called as hemoglobin S and increased sensitivity to infections. It affects different organ with alteration in joint, bone, gastrointestinal tract, kidney etc. which ultimately increase mortality and morbidity rate in affected group of patients. Osteoarticular manifestation commonly seen in SCD marked as arthritis, osteonecrosis and osteomyelitis. Case on sickle cell with arthritis were rarely reported. In this case patient presented with sickle cell disease with left elbow monoarthritis which was treated accordingly and discharged with follow-up advice to family member with plan for to start hydroxyurea totally based on patient condition.

Keywords: Sickle cell disease, vaso-occlusive crises, osteonecrosis, osteomyelitis, arthritis.

I. INTRODUCTION
SCD is inherited lifelong blood disorder, named as such due to its resemblance in the shape of a sickle. Homozygosity and heterozygosity are the two form of mutation happen commonly in the sickle which defines it as either sickle HBSS “sickle cell disease” or HBAS “sickle cell trait”. Its shape tends to lodge in various organs and joints which result in inflammation and pain at accumulation site. The most perennial issue that requires hospitalization in SCD patients is painful vaso-occlusive crises, arthritis, osteomyelitis and osteonecrosis.

In SCD arthritis generally associated with a polyarticular and symmetric form with an inclination for large joints and lower extremities, mostly last for less than one week. When assessing for joint issues in SCD, inpatient X-ray helps in identification of space between the joints and bones, synovitis, bone erosion and other types associated condition with SCD. Commonly associated entities in SCD are avascular necrosis, osteomyelitis, bone infarcts, and septic arthritis. Synovial effusion is responsible for 20% of acute attacks. Acute septic arthritis happen rarely in SCD and often portray as monoarthritis.

Sickled RBC in micro capillary cause enormous and unendurable pain, sometime even it does not manage with painkillers. No other abnormal Haemoglobin condition has such tendency to lead patient life miserable except patient suffering from SCD. Patient and members of the family face a different challenge and concern while managing the disease with health care providers. Various organization, international and national collaboration, public health works, health care planning experts all are working with an objective of more appropriate and precise therapy, proper management guideline, cover socio-economic, behavioral aspects and counselling available to both developed and low income developing countries for better patient care and management of SCD. Currently, 3 main strategies available for management of SCD and its complication are blood transfusion, bone marrow transplantation and hydroxyurea administration.

CASE REPORT
A 3 year old male child presented with complaint of difficulty in breathing along with cough for 15 days and left elbow joint swelling for 2 days. There is no history of any trauma, septic arthritis, gonococcal arthritis, Poly arthritis, tubercular arthritis and hemarthrosis. Patient was resident of Madhya Pradesh and also had a history of 2 unit blood transfusion. Systemic examination was performed which shows that pallor was present, Patient was conscious to time place and person, abdomen was distended, non-tender, B/L wheeze present and S1, S2 sound was heard. Based on history taking and clinical manifestation, the patient provisional diagnosis was acute respiratory infection with respiratory distress with swelling in left elbow joint query anemia. Show in fig 1.

Fig 1: patient with left elbow swelling
Symptomatic treatment was provided to ease patient symptoms while further investigation was performed. Treatment include O₂ inhalation, IV infusion 5% GNS + KCL, inj Augmentin, inj monocef, inj febrinil and syrup multivitamin. Investigations revealed that Hb (7.7 g/dl), lymphocytes (20%), ESR (32 1st hr mm) and Neutrophils (75%). PBS Report shows – moderate dimorphic...
anemia with neutrophil leukocytosis (Target cells seen occasionally holy leaf seen.) C- reactive protein test – was positive (> 24 mg/lit < 48 mg/lit). X RAY of elbow – left elbow monoarthritis. Shown in fig 2.

Fig 2: left elbow monoarthritis

HB – Electrophoresis – HB-P3 – 2.7%, HB-AO – 32.0% a HB-F – 12.7%, HB-S-43.0%, HB-A2 – (H) 4.8% Consistent with thalassemia trait with heterozygous for Haemoglobin S (sickle cell), SERUM IRON - Low 17.1 ug/dl, Transferrin serum iron saturation - Low 7.09%, UIBC – 224.2 ug/dl, SERUM T.I.B.C low 241.3 ug/dl, Serum ferritin - 96.53 ng/ml, USG W/A normal and Mutation analysis did not done due to financial issues. Based on the investigation result, final diagnosis was sickle cell disease with left elbow monoarthritis. Finally the treatment was continued for the duration of 10 days and 2 PRBC transfusion was done with continuous monitoring. Discharge with follow-up advice to patient family members and also plan for hydroxyurea administration on follow up on the basis of patient condition and need.

DISCUSSION
A most common misconception about SCD is that it only affects the people of low socioeconomic status and certain races, however, it affects the person of any races and ethnicity worldwide. In Indian tribal population, SCD prevalence varies from 1-40%. Highest load of homo and heterozygosity is seen in Madhya Pradesh. It consists of 45 districts out of which 27 comes under the sickle cell zone varies from 10 to 33%. The chances of having disease on birth in a newborn are much high due to the presence of homozygotes. [6, 7] Patient with the disease is also highly vulnerable to other infections. Presence of SS and AS in parents define whether the baby is at risk of SS or AS. If one parent is having AS and one is having SS there is 50 – 50% chance of having either SS or AS. When both parents carry AS then there is 25% chance of developing SS in a child. [8, 9]

Sickle Hb polymerization works as a catalyst for vaso-occlusion. The certain factor is critically involved in the pathophysiology of sickling listed as WBC, vascular endothelium, abnormality in coagulation and damage to RBC results in a vasculopathy and hemolytic anemia. In children with SCD ischemic stroke is one of the major threat which includes common complication such as aplastic episodes due to infection, acute chest syndrome, sequestration crisis and febrile events. [10]

While in adult SCD it present with unique complication including priapism, pulmonary hypertension, delayed sexual development, avascular necrosis of hip and gallstones. Treatment approach in SCD based on patient condition. Some patient needs extensive care while some need routine check-up depending upon its variable clinical manifestation and progression. A multidisciplinary team approach is needed for a patient with an extensive facility care center. [11, 12]

In this case report, patient was admitted to the hospital with a complaint of respiratory distress and swelling in the left elbow. The investigation was performed for definitive diagnosis and investigation conclude that the patient was having SCD with left elbow monoarthritis. Treatment was planned and provided to a patient with counselling to family members regarding the patient condition and disease prognosis. The patient was discharged with discharge medication and follow up advice.

CONCLUSION
Sickle cell disease requires timely, proper history taking and diagnostic approach for better patient care and management of a disease. The treatment option available for SCD is mostly symptomatic. Different osteoarticular involvement, generally one of the common reason for a patient to go to the emergency department. Mechanism of pathophysiological complication which is commonly developed in SCD needs to be studied for proposing more precise and accurate guideline for treatment. Parents should be given genetic counselling and support. Presently patient is maintaining normal routine life with proper clinical management.

ACKNOWLEDGMENT
The author acknowledging honorable faculty members and staff of paediatrics department at NIMS Institute of Medical Science and Research, Jaipur, Rajasthan. Author would also like to thanks and appreciate the patient family members and attender for their contribution and permission.

CONFLICTS OF INTEREST
The author declared that there are no conflicts of interest related to this study.

AUTHORS’ FUNDING
No any source of funding.
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