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Case Report On 21-Year-Old Male with Sickle-Cell Disease and Growth Retardation.

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ABSTRACT
Introduction: Due to abnormal haemoglobin synthesis, sickle cell disease is often called autoimmune hemolytic anaemia. Red blood cells sickle due to faulty hemoglobin, which causes a vaso-occlusive crisis.
Case presentation: A 21-year-old male was admitted to a tertiary care hospital with a known case of sickle-cell disease and growth retardation. The patient was apparently alright 13 days back when he complained of a hematemesis episode of yellowish discolorations of the body 13 days. Acute onset of pain in bones and joints for the last seven days and weakness,fatigue for the last 12 years and recurrent attacks of pallor. The patient had known complaints of portal hypertension with decompressed autoimmune liver hepatitis, sickle cell anemia, and AS-pattern with auto splenomegaly. Recently there was no history of cough, cold, nausea, vomiting, abdominal pain, chest pain, palpitation, and breathlessness.no other associated illnesses, i.e. hypertension, diabetes mellitus, and tuberculosis. He was admitted to the hospital with the above mentionedcomplaints. On admission, his weight was only 20 kg. The patient had a family history of sickle cell disease father and mother are AS patterns, and the elder sister also has AS patterns. The patient's physical examination and all routine laboratory and radiological investigations were done. The patient's Hemoglobin was 5.8gm%. As per the doctor's order, the patient was kept under close observation, and medical treatment was started. The doctor advised them to liver transplantation. The patient's and his relatives' psychological counseling were done. But the patient is not willing to stay at the hospital. The patient was stable at present, so the patient was being discharged.
Conclusion: Early diagnosis and treatment can prevent the severity of the disease and its complications.

Keywords: Sickle cell disease, hematemesis, weakness, growth retardation.

INTRODUCTION:

Sickle cell disease or sickle cell anemia is a red blood cell disorder. Round and healthy red blood cells travel via small blood channels to deliver oxygen to every part of the body. Sickle cell disease occurs due todefective haemoglobin formation. In sickling of red blood cells occur due to defective hemoglobin, which leads to vaso-occlusive crisis. In this condition, red blood cells become a sickle shape. The alternative complement pathway is defectively activated in sickle cell disease (S.C.D.) patients, which raises the risk of infection and may predispose them to autoimmune illness[1].

Various tribal ethnic groups in India carry the sickle gene, and the proportion of heterozygotes ranges from 1 to 40%.After birth, newborn screening programs for sickle cell disease have recently been seen in Chattisgarh, Maharashtra, Gujarat, and Odisha; monitoring these birth cohorts will help to understand the natural history of sickle cell disease in India.Indian tribal families accepted prenatal diagnosis. The National Rural Health Mission and the Indian Council of Medical Research are implementing outreach programs in various States. For better monitoring and control of the spread of the disease.[4]

Sickle cell anemia is known as hemolytic anaemia of this condition. Hemolytic anemia was the first symptom this patient had. This instance was identified by using haemoglobin electrophoresis and a peripheral smear, antibiotics, steroids, Blood transfusion, loop diuretics, and oxygen inhalation; all treatments helped this patient to recover.Sickle cell patients are more prone to recurrent bacterial infections, primarily pneumococcal infection. Hydroxyurea is

helpful to advance therapy in sickle cell anemia, but those patients on hydroxyurea experienced repeated episodes of acute chest pain[2].

Growth retardation seen in Sickle cell disease,delayed sexual development, and being underweight during childhood and adolescence. In adolescents with sickle cell disease, Rhodes et al. mentioned that growth delays during puberty are independently correlated with a decline in Hemoglobin in total energy consumption. And increased total energy expenditure.

Adolescents with sickle cell disease experienced puberty more slowly than healthy control children, according to Rhodes et al. The height of affected pubertal males decreased with time, and they grew shorter than their natural counterparts. However, they did not vary in terms of annual weight gains[3].

Sickle cell disease affects several systems in the body. The habitus of children with Sickle cell disease is a result of the involvement of the musculoskeletal system. The changes in body structure; are small for age due to growth retardation, increased anteroposterior diameter of the chest, and resultant chronic hypoxemia of the bone marrow leading to bone marrow hyperplasia.

CASE PRESENTATION:

The 21-year-old male was admitted to a tertiary care hospital with a known case of sickle-cell disease and growth retardation. The patient was apparently alright 13 days back when he had a complaint of hematemesis episode yellowish discoloration of the body 13 days. Since last seven days have seen a sudden start of bone and joint pain, while the past 12 years have seen repeated episodes of pallor, weakness, and fatigue. The patient had known complaints of portal hypertension with decompressed autoimmune liver hepatitis, sickle cell anemia, and AS-pattern with auto splenomegaly. No history of cough, cold, nausea, vomiting, abdominal pain, chest pain, palpitation, and breathlessness. No other associated illnesses, i.e. hypertension, diabetes mellitus, or tuberculosis. The client was admitted to a tertiary care hospital with the complaints mentioned above.On admission, patient's weight was only 20 kg. The patient had a family history of sickle cell disease father and mother are AS patterns, and the elder sister also has AS patterns.

On physical examination: patient's body temperature was -101^{0}F, pulse-120 beats/min, and respiration- 34 breaths/min, blood pressure was -124/82 mm of Hg, mild edema and pallor present, Cardiovascular system –normal S1S2 heart sounds present, no murmur, Respiratory system - bilateral crepitations, abdomen - hepatosplenomegaly was found. Electrocardiography - sinus tachycardia present. Central nervous system: during examination patient was fully conscious and oriented to time, place and person. Bowel sounds were present, and it was normal.

This male, age 21, has already had sickle cell anaemia since he was a young child. Short stature, shunted growth. The patient had complained of recurrent jaundice since three years of age. The patient had a history of major prominent presented with hematemesis and malaria. Recurrent jaundice with decompensated chronic liver disease. Currently, he was admitted to the hospital for a liver biopsy. Given recurrent jaundice and hemolytic anemia, an ASMA test was done, i.e.an anti-smooth muscle antibody blood test weekly positive, IgG autoimmune hepatitis. His simplified score was 8. Hence he was diagnosed with definitive autoimmune hepatitis—positive and liver reports suggestive of gamma-globin 1 to 1.5 times of UNL. The ultrasonography report showed cirrhosis. Investigated for etiology- viruses negative. The patient had large esophageal varices – presentation as massive variceal bleed-EVL has done.

All Routine investigationswere done; the report attached ultrasonography showed urinary wall thickened 6mm, suggested of cystitisparenchymal disease. Laboratory reports revealed that Hemoglobin was 5.6gm%, MCHC-34.4, MCV-139.1, MCH-47.9, Total Red blood cells count was 1.16, total White blood cells count- 5400, Total Platelet Count- 1.45,Hematocrit-16.2.R.B.C.s- Normocytic Hypochromic with mild aniso-poikilocytosis showing few microcytic and pencil cells. Platelets –on the lower side of the normal range. No haemo-parasites were seen. Kidney function test revealed that, urea 63mg/dl, creatinine 0.9 mg/dl, sodium 136 mEq/l, potassium 2.5 mEq/l. The liver function test report revealed alkaline phosphatase 133, SGPT -22, SGOT 58, total protein 8.0, total bilirubin was 32.1, bilirubin conjugated 11.0, and bilirubin unconjugated 21.0, globulin calculated parameter 4.9.

Therapeutic management in hospital: Inj pan 40 mg IV BD x 4 days, Syp sucralfate 2 tsp TDS, tab zin OD, the pup with o2 at 2l/min, Inj KCL 40 meq in 500ml over 4 hours**.** Treatment on discharge: Inj pan 40 mg IV BD x 4 days, syrup. Sucralfate 2 tsp TDS, Tab zin OD, the pup with 02 at 2l/min, Inj KCL 40 meq in 500ml over 4 hours.

Nursing management: Advised to patient's caregivers to givea diet high in vegetables, fruits, legumes, and whole grains, and usually provide an adequate amount of vitamins and macro and micronutrients to support the health Patient. The patient's family members were so worried about his illness that sometimes the patient showed anger and

became aggressive in his behaviors, and related diseases he had so many queries and doubts. That was clear by health care personnel. The patient and his family members' psychological counselling was done. The doctor advised them to liver transplantation. But the patient was not willing to stay in the hospital. The patient was stable at present**;** hence the patient was being discharged.

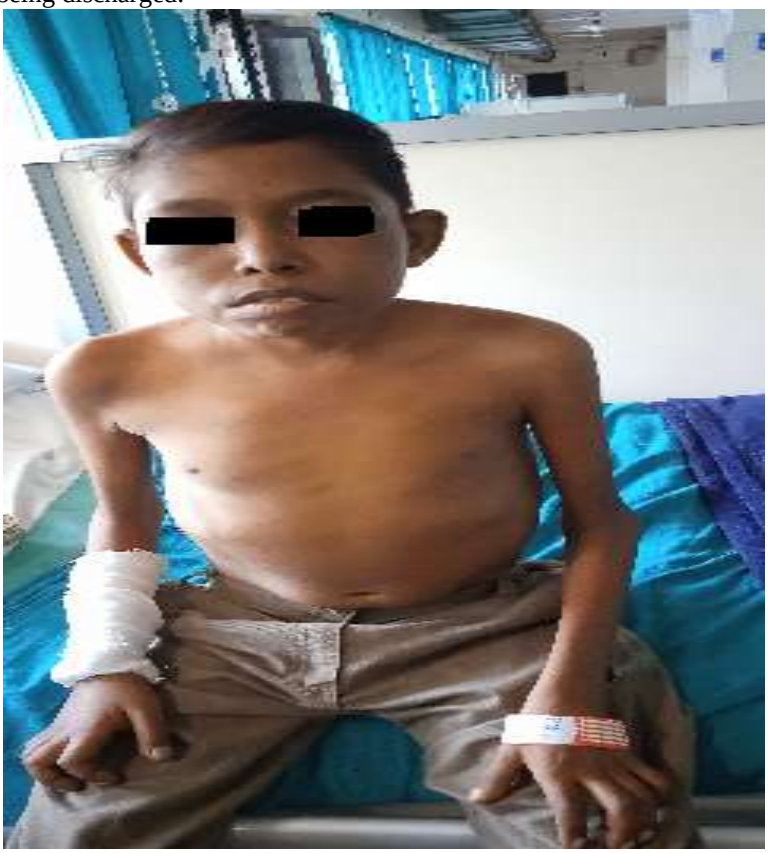

Figure 1: A Clinical Image Of A 21-Year-Old Male With Sickle-Cell Disease And Growth Retardation.

DISCUSSION:

We report a case on a male patient, age 21, with a diagnosed sickle-cell illness who was also diagnosed with autoimmune hepatitis, auto-splenectomy with an AS pattern, and growth retardation**.** He had a history of recurrent jaundice for 12 years. On admission, his weight was only 20 kg. There was a family history of sickle cell disease among the patient's family.His father and mother both are AS patterns, and his elder sister also has AS patterns. After admission patient's physical examination was done, all routine laboratory investigations were done, and on report patient's Hemoglobin was 5.6gm%. Therapeutic management was done. The doctor advised them to liver transplantation. But the patient was not willing to stay in the hospital. The patient was stable at present, so the patient was being discharged.[5-14]

This present patient was already diagnosed with sickle cell disease since childhood: short stature, shunted growth. The patient had complained of recurrent jaundice since three years of age. The patient had a history of major prominent presented with hematemesis—recurrent jaundice with decompensated chronic liver disease. In the past, patients had received a treatment tab. Prednisolone 30 mg, tab.Pantoprazole 20 mg, tab.Ciplar 40 mg, tab.Ursodexocholic acid 300mg and tab.kenadon, Inj vitamin K. tab. Vitamin B_{12}, tab. Folic acid and radiological investigations were done.[14-21]

In this study researcher mentioned about, assess the risk factors regarding growth retardation among children who have sickle cell anemia. This research study aimed to evaluate theincidence and, secondly, to identify the risk factors regarding growth retardation and sickle cell anaemia. Study findings compared to previous research studies reported in the literature. As endocrine dysfunction and other metabolic dysfunction, hematological condition, and patients with sickle cell disease may experience growth failure because of their nutritional status[21-26].

CONCLUSION:

The present patient presented with the mentioned health problems and was admitted to a tertiary care facility; afteradmission, the patient's health checkup and all routine investigations were done. And the medical treatment was started, but the patient did not willing to stay at the hospital. The patient was stable at present, so the patient was being discharged. Children with sickle cell disease have growth retardation and get worse with age. This retardation is associated with the male gender, low socioeconomic status, and severe or uncontrolled illness symptoms.

ETHICAL APPROVAL: Not applicable

PATIENT INFORM CONSENT: While preparing a case report for publication patient's informed consent has been taken.

CONFLICT OF INTEREST: The Author declares that there are no conflicts of interest.

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