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A RARE CASE REPORT: HODGKINS LYMPHOMA WITH SICKLE CELL DISEASE IN AN ADULT PATIENT

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ABSTRACT

Hematological malignancy has rarely been reported in adults with sickle cell disease. There are very few cases of patients with sickle cell disease who developed Hemato-oncological malignancy including myeloid and lymphoid conditions. We here report the case of hodgkins lymphoma with sickle cell disease in an adult from central India.

KEYWORDS:

Sickle cell disease, hodgkins lymphoma

Introduction:

James Herrick, a physician first described the characteristic sickle shaped red cells in a medical student from Grenada in 1910. Linus Pauling and his colleagues showed that sickle haemoglobin (HbS) had an altered electrophoretic mobility and they were the first to define it as a molecular disease in 1949. A few years later in 1957, Vernon Ingram discovered that sickle haemoglobin resulted from a single amino acid substitution in the haemoglobin molecule[1].

Sickle cell disease is a global public health problem. According to CDC statistics data , it is estimated that sickle cell disease affects approximately 100,000 Americans. It occurs among about 1 out of every 365 Black or African-American births and 1 out of every 16,300 Hispanic-American births. About 1 in 13 Black or African-American babies is born with sickle cell trait (SCT)[2].

Sickle haemoglobin is highly prevalent among the tribal of central, southern and western India [3,4] with variable frequency ranging from 10-23%. Increased prevalence is also reported in the non tribal communities of the above said areas. Amongst all states Maharashtra, Madhya Pradesh, & Tamilnadu have higher prevalence of this disease. Central India region is a focus of sickle cell disorder and the prevalence in Vidarbha region of Maharashtra ranges between 4-40% with average sickle cell gene frequency being 4.3%[5].

This disease has variable clinical presentation and most of the Indian patients remain asymptomatic for longer periods due to higher levels of HbF[6]

Approximately 20 percent of children with SCD die by the age of two [7] Sickle cell disease (SCD), which causes a wide range of severe and even life threatening consequences, is caused by a single misspelling in the DNA instructions for hemoglobin, a protein vital for carrying oxygen in the blood. As a result of this mutation, individuals with SCD experience lifelong complications including anemia, infections, stroke, tissue damage, organ failure, intense painful episodes, and premature death. These debilitating symptoms and the complex treatment needs of people living with SCD often limit their education, career opportunities, and quality of life . Many complications of SCD can make every stage of life extremely challenging for individuals with the disease[8]. Patient with sickle cell disease or trait can develop malignancy of lymphoid or myeloid origin[9] but is considered to be very rare[10]. Here we report an uncommon case of sickle cell disease with hodgkins lymphoma.

Case Presentation:

A 38 years old male known case of homozygous sickle cell anaemia, Mahar (SC) by caste residing at Gadchiroli district of Maharashtra state presented with low grade fever and neck swelling since 2 months at tertiary care hospital of central India. On examination , pallor was present. Liver was palpable. Right cervical Lymph Nodes palpable, matted 1cm.

Follwing Investigations were done:

- Hb:6.2gm%,TLC:6000,DLC:N:70%,L:26%,E2%M2%.
- LDH:425,LFT/RFT: WNL,Alk Phosphatase:354IU
- X Ray: Bilateral Hilar Lymphadenopathy present
- Bone Marrow shows Microcytic Hypochromic anaemia
- CT Abdomen reveals Multiple intraabdominal and thorasic lymphadenopathy, hepatosplenomegaly and presence of splenic calcifications.
- USG abdomen gave evidence of Fatty liver, hepatosplenomegaly and gall stones.

Histopathological examination Report:

- Gross: 5 firm grey white colored lymph nodes
  2cm ×0.5 cm to 1cm ×1 cm
- Microscopic: Complete loss of lymph node architecture with intact capsule ,showing mixed cellularity against a background of mature lymphocytes ,many pleomorphic histiocyte like cells with large vesicular nuclei seen , many Reed Sternberg cells having binucleate cells with each nucleus forming mirror image of each other
- Nuclei are vesicular with prominent eosinophilic central nucleolus.
- Many multinucleated giant cells seen
- Areas of necrosis also seen.
- OPINION: Lymphoreticular Malignancy (Hodgkins Disease)
- Clinical Diagnosis: Hodgkins Lymphoma(3b)
- Patient received 4 rounds of chemotherapy drugs containing Adriamycin, Bleomycin , Vinblastin, Dacarbazine.

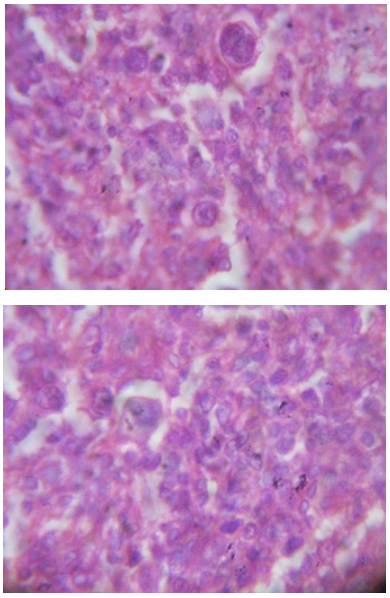

Fig 1&2: Histopathological finding showing binucleated reed sterberg cell

Discussion:

On the basis of a single institution study, the cancer incidence among patients with sickle cell disease has been reported to be 1.74 cases per 1,000 patient-years(11) .William H. Schultz et al conducted a study in Calornia to know incidence of malignancy in a patient with SCD in that 3 cases of lymphoma was reported(12) Hematological malignancies are uncommon in sickle cell disease.

B.J.Brown reported hodgkin lymphoma in a 7 year old male child with sickle cell anemia (13)

Stricker et al described four sickle cell patients (two with hemoglobin SC, two with hemoglobin SS) who developed hematologic malignancy (acute myeloblastic leukemia, multiple myeloma, malignant histiocytosis, and Hodgkin's disease) (14) . Santosh G. Rathod et al reported a rare case of chronic myeloid leukemia with sickle cell disease from Aurangabad Maharashtra India. Other reported hematological malignancy occurring in Sickle cell disease patients are, acute myeloid leukemia (AML), acute lymphoblastic leukemia, multiple myeloma, malignant histiocytosis, B and T-cell non-Hodgkin's lymphomas -T cell lymphoma and chronic lymphocytic leukemia, hairy cell leukemia.(9,14,15,16). Risk factors for hematological malignancy in sickle cell disease put foreword; which includes, infection like HIV virus, hepatitis C virus, persistent transfusion related immunomodulation, stem-cell transplantation, and chemotherapeutic agent like hydroxyurea used in treatment of sickle cell disease (9,17,18) .

Conclusion:

Advances in medical therapy, screening and patient education will prolong the life span and can improve quality of life of many patients with sickle cell disease. The average life expectancy in patients with sickle cell disease has improved significantly. We need more epidemiological and cytogenetic studies in the adult sickle cell population to assess the incidence and causative factors of hematological malignancy in these patient.

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