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Clinical Manifestation in Sickle Cell Anemia: A Study in Chhattisgarh Institute of Medical Sciences (CIMS) Bilaspur , Chhattisgarh , India.

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Abstract:
Objective: *Sickle cell Anemia(SCA) is an inherited disorder that affects red blood cells. In this study we retrospectively analyzed 104 patients, who presented to the medicine OPD at CIMS, Chhattisgarh, India. Clinical parameters and manifestation were studied in order to know pathophysiological complication going on in SCA.*
Material methods: *Retrospective analysis of data was done in patients of sickle cell disease referred to CIMS Bilaspur Chhattisgarh for a period of 6 months in the year 2010.*
Results: *Total subjects enrolled were 104 of sickle cell anemia patients. 26.0% were in pediatric age group and 74.0% comprised of adult populations. 51.9% males and 48.1% females contributed in our study. Clinical manifestations was gathered and filled in data sheets it was observed that 19% patients were severely anemic , 62% were moderately anemic , 12% had mild anemia. Fever was observed in 96.2% of patients. Occurrence of pain episodes were in 86.5%, 80.7% experienced recurrent tiredness, swelling of abdomen was noted in 30.6 %, 36.5% experienced unusual headache. From the data sheet it was found that 66.3% of sickle cell anemia suffered from Jaundice. Growth retardation was observed in 28.8%, recurrence occurrence of pain episode was found in 60.0%.Acute chest syndrome in 37.5% patients, Hand foot syndrome 21.1%.Chest infection was found in 19.2%, 82.6% was found to have vaso-occlusive crisis, malarial fever was in 0.4% patients, cholilithiasis was detected in 0.3% patients, spleenomegaly was found in 42.3% patients, hepatomegaly was found in 50.0%, hematuria was recorded in 0.5% patients, leg ulcers were found in 0.6% patients, 42.2% had history of multiple blood transfusion.*
Conclusion: *Thus sickle cell anemia in district Bilaspur and catchment areas shows diverse clinical manifestations in variable percentage. Thus it is proposed to manage the disease properly as soon as it is detected and to have a definite approach comprising of public awareness to reduce the stigma of Sickle cell disease.*
Keywords: *Sickle cell anemia (SCA), Sickle cell disease, clinical manifestation.*

I. Introduction

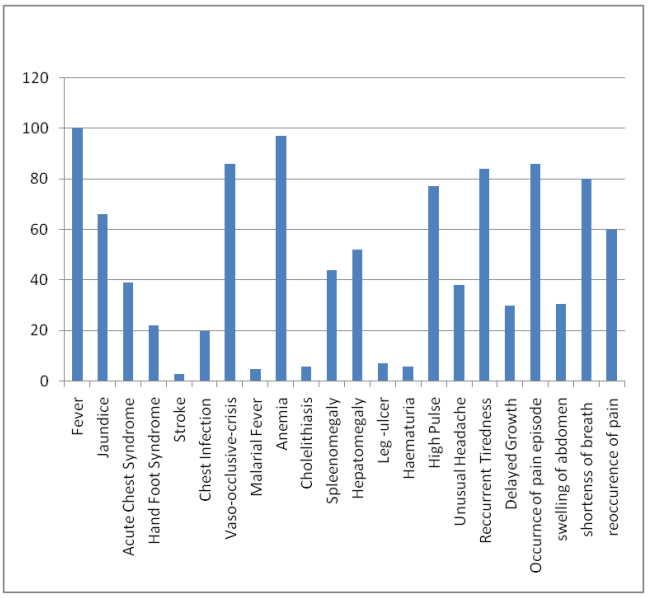
Sickle cell disease is a common single gene disorder, 50% of world population resides in India[1] . Sickle cell anemia is caused due to substitution of Valine for glutamic acid at 6th position of beta globin of haemoglobin[2,3]. This mutated and defective hemoglobin is less soluble and after deoxygenation undergo polymerization causing distortion of shape of (RBC) red blood cell[4]. The pattern of inheritance of this disorder is recessive. SCA carrier (trait) are asymptomatic generally are not aware of diseased gene they carry. The disease is found predominantly among high risk communities belonging to Scheduled Tribe Scheduled caste and other backward classes in Chhattisgarh. Approximately 3.2million populations belong to high risk community in central India, there rare 250,000 children worldwide everywhere. Most of them die before they attain the age of 2 years, due to multiple stroke[6,7]. Life expectancy of SCA patients in less. SCA patients die due to complications like Pain, acute chest syndrome and stroke[8].Hence this study was undertaken at CIMS Bilaspur Chhattisgarh, in order to observe the clinical manifestations and to know the pathophysiological complications going on in sickle cell anemia.

II. Material methods

This is retrospective, observational study carried out on sickle cell anemia patients attended in OPD. Most of the patients ,about 51% attended were from Bilaspur district and rest were from Durg (12%), Janjgir (11%) , Rajnandgaon(3%), Raigarh (6%), Korba (7%),sarguja (2%), Kawardha (4%), Korea(1%), and jaspur (1%). Sample size was 104 sickle cell anemia patients. The mean age of selected patients were 25.4%. The study was approved by ethical committee and college authority. Patients health history was undertaken to understand the various complication going on the body of patients. Socioeconomic status of the patients were

noted. Some of the parameters and some of the complications as illustrated in figure 1. A statistical analysis was done by Pearson chi square using SPSS 11.5 version software.

Illustration 1



Clinical complication presented by sickle cell anemic patients. [y-axis shows percentage].

III. Results:

Clinical manifestations such as joints and chest pain, shortness of breath , swelling of abdomen were observed in sickle cell anemia. total subjects enrolled were 104 of sickle cell anemia patients. 26.0% were in pediatric age group and 74.0% comprised of adult populations. 51.9% males and 48.1% females contributed in our study. Clinical manifestations was gathered and filled in data sheets it was observed that 19% patients were severely anemic , 62% were moderately anemic , 12% had mild anemia. Fever was observed in 96.2% of patients. Occurrence of pain episodes were in 86.5%, 80.7% experienced recurrent tiredness, swelling of abdomen was noted in 30.6 %, 36.5% experienced unusual headache. From the data sheet it was found that 66.3% of sickle cell anemia suffered from Jaundice. Growth retardation was observed in 28.8%, recurrence occurrence of pain episode was found in 60.0%.Acute chest syndrome in 37.5% patients, Hand foot syndrome 21.1%.Chest infection was found in 19.2%, 82.6% was found to have vaso-occlusive crisis, , cholilithiasis was detected in 0.3% patients, spleenomegaly was found in 42.3% patients, hepatomegaly was found in 50.0%, hematuria was recorded in 0.5% patients, leg ulcers were found in 0.6% patients,malarial fever was in 0.4% patients out of which patient (n=1), had Plasmodium falciparum positive and rest were vivax positive. Patient (n=1) had HIV positive 42.2% had history of multiple blood transfusion.

IV. Discussion

The present study demonstrates, majority of patients suffered from anemia, out of which 20.0% had severe anemia (hemoglobin <5gm), 65% of patients had moderate anemia, (hemoglobin >5-8gm) and 12% had mild anemia (hemoglobin >8-11gm), severe anemia was observed in pediatric age group as compared to adults , probably because of repeated infections by pathogens , inadequate nutrition or excessive need of growth. These findings were observed by other workers[9,10,11.]Fever was experienced in 66.3% was less as compared to study conducted by Wankhede.V et al[12]. Recurrent tiredness was noticed in 80.7% of cases, which was predominant finding. Recurrence of pain was observed in approximately half of patients i.e. 55.5%. swelling of abdomen was found in 30.6% of patients, Malarial fever was noted in 0.4% out of which 1 patient (n=1) had Plasmodium falciparum and rest had Plasmodium vivax positive. HIV was positive in (n=1) , this patient had history of multiple blood transfusion. Some common mode of clinical presentation of SCA shown in figure 1.Clinical manifestation in Sickle cell anemia were diversed and it is not well understood and explained by single gene mutation. Variability regarding symptoms is observed in the frequency and intensity of painful vaso-occlusive crises and also in the degree of organ dysfunction. Pathophysiology appears because of combined effects of hemolysis and vaso-occlusion. Hemoglobin picks up and releases oxygen repeatedly and undergoes polymerization and depolymerization leading to hemolysis. As a result free hemoglobin uses significant amount of nitric oxide (NO) that results in abnormal regulation in the vascular homeostasis[13,14]. In SCA, important cause of morbidity and mortality is ACS (Acute Chest Syndrome). In our study 37.5% patient had ACS. ACS which

found to occur in 45% of patients and recurred in 80% afflicted patients[15,16]. The chief pathologic incidence in ACS is vaso-occlusion, the etiology is multifactorial. Abnormal adherence of sickle RBCs, WBCs and/or platelets to the vascular endothelium is one of the phenomenons causing vaso-occlusion. It is not clearly understood about factor leading to cellular adhesion and vascular damage. Oxidizing molecules for example O2-, H2O2, •OH radical and ONOO-are produced in large number during periods of reperfusion[17]. These compounds have ability to activate second messengers that causes up regulation of endothelial adhesion molecules. Molecules such as vascular cell adhesion molecule (VCAM)-1 and intercellular adhesion molecule (ICAM)-1, facilitate binding of sickle RBCs and WBCs to the vascular endothelium and thus may be responsible for vaso-occlusion[18-21]. In our patient vaso-occlusive crises was noted in 82.6%, this percentage was reasonably high. Further, oxygen-related species directly can damage endothelium by per-oxidation of the lipid membrane and/or DNA fragmentation, causing cellular apoptosis**.** SCD patients are subjected to great oxidative stress, mainly during vaso-occlusive crises (VOCs) and ACS. This is represented by increased levels of oxygen radical and leukocyte adherence and emigration[19-22]. In our patients stroke was experiencd in 0.2% patients of SCA. Risk of stroke to SCA patient is 221 –fold higher than healthy child[24]. Within the renal tubular epithelium ONOO- are formed with associated cellular apoptosis[23].In our case reported cases of headache 36.5%. Niebanck et al[22]. reported cases of headache in sickle cell anemic patients. Many factors may involve in the symptoms of headache in sickle cell anemia and linked to migraine, bone marrow hyperplasia, OSA, or cerebral vessel stenosis[29]. In sickle cell anemia, tissue ischemia resulting in acute and multi-organ dysfunction is caused by intermittent episodes of vascular occlusion[30] which is explained by chronic inflammation[19,22,31] and ischemia-reperfusion injury[24,25,26,27]. Neutrophils have significant function in tissue damage . Adhesion of lymphocytes and monocytes to the endothelium sickled induced by red blood cells may add to the pathogenesis of vascular occlusion . The observations of clinical manifestations and parameter are in support of above mentioned studies.

V. Conclusion:

Sickle cell patients of district of Bilaspur and near catchment areas are under great stress. Among the SS and AS subjects 32% belong to backward caste, a comparatively higher number as against scheduled caste and tribe which were about 18 and 15% respectively. This data indicate that sickle cell gene is predominant among the backward caste and schedule caste population in adjoining tribal population at Bilaspur and surrounding district. It is contemplated to understand the natural history of haemolytic anaemia, identify homozygotes follow them regularly and provide immediate facility and earlier intervention to manage the complication in sickle cell anaemia patient. There is a need to carry out larger scale study. Counselling, awareness and effective intervention by different government agencies will definitely reduce the stigma attach to this disorder. If remain ignored it will result in extinction of some castes and tribal from this state.

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