Clinical Differences in Severity between Pediatric and Adult Sickle Cell Anemia Patients in Wayanad District of Kerala

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Abstract: Sickle cell disease is a multisystem disease with acute illness and progressive organ damage with severe monogenic disorders worldwide. SCD can be accompanied by numerous physical symptoms and medical complications such as recurrent pain, anaemia, low exercise tolerance, splenic sequestration, susceptibility to infections, lung problems, growth delay, late onset of puberty, strokes, priapism, enuresis, and decreased life expectancy. Physiological complications for children include pneumococcal infections, meningitis, osteomyelitis, cerebral vascular infarction, and enuresis. To determine the severity between children and adult sickle cell anemia patients a study was conducted among the patients attending Swami Vivekananda Mission Hospital, Muttil, Wayanad district of Kerala and also from the mobile clinics. In the study it was found that the clinical severity is more in adults than children,

KEYWORDS: SCD, anemia, sickling test, electrophoresis, Clinical finding, SPSS

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INTRODUCTION

Sickle cell disease is a multisystem disease, associated with episodes of acute illness and progressive organ damage, and is one of the most common severe monogenic disorders worldwide [1]. These disorders are found among many different ethnic groups including those of Mediterranean, Carribean, South and Central American, East Indian and Arab descent [2]. Herrick first described the characteristics sickle-shaped erythrocytes in 1910[3]. Pauling and Colleagues identified electrophoretic abnormalities in sickle hemoglobin (HbS) and coined the term "molecular disease" in 1949[4] . Persons with SCD have a preponderance of red blood cells that become hardened, brittle, and sickle-shaped when they release oxygen. This hardening and sickling of haemoglobin may result in vaso-occlusion, swelling, and pain episodes. These haemoglobins are fragile and do not live as long as normal red blood cells. Anemia is the consequence, then, not of cellular iron deficiency but rather of the abbreviated life span of the sickle haemoglobin. SCD can be accompanied by numerous physical symptoms and medical complications such as recurrent pain, anaemia, low exercise tolerance, splenic sequestration, susceptibility to infections, lung problems, growth delay, late onset of puberty, strokes, priaprism, enuresis, and decreased life expectancy [5] . Physiological complications for children include pneumococcal infections, meningitis, osteomyelitis, cerebral vascular infarction, and enuresis [6] The recurrent pain caused by the disease can interfere with many aspects of the patient's life including education, employment, and psychosocial development. In sickle cell anemia, a lower than normal number of red blood cells occurs because sickle cells do not last very long. Sickle cells die faster than normal red blood cells, usually after only about 10 to 20 days [2] . The bone marrow cannot make new red blood cells fast enough to replace the dying ones. This result in anemia.

METHODOLOGY

Objective – to determine the severity between children and adult sickle cell anemia patients.

Study population – patients attending Swami Vivekananda Mission Hospital, Muttil, Wayanad district of Kerala and also from the mobile clinics were screened for sickle cell anemia by sickling test followed by confirmation with hemoglobin (Hb) electrophoresis. This was a cross sectional study between 2 group of patients, 100 patients in each group (group1includes pediatric patients and group2 contains adult patients). Only homozygous patients were included. Among children the age group is within 6 months to 14 years and adults the age group is within 15 to 40 years. Clinical and hematological data were obtained from all the patients. Based on the findings present in proforma the sign and symptoms between these two groups were correlated and the severity of the disease was found out. The data were analyzed statistically (using SPSS) with chi-square test.

ETHICAL CLEARANCE

The study topic along with case proforma was presented before the Ethical committee of G.A.C, Pariyaram, Kannur. The objectives, methodology and expected outcome of the present study were explained in the committee. The committee after discussion gave the ethical clearance to conduct the study.

CLINICAL FINDINGS

PAIN	GROUP1 Number	GROUP2 Number	P value
Chest + limb	12	4	
Chest + joint	8	12	
Chest+ abdomen	2	–	
Limb + joint	4	2	>0.1
Limb + abdomen	4	–	
Joint + abdomen	8	2	
Chest + limb + joint	4	24	
Chest + joint + abdomen	22	16	
Chest + limb + joint + abdomen	36	40	
TOTAL	100	100	

FEVER	GROUP1 Number	GROUP2 Number	p value
In every month	16	20	
4 times in year	38	34	
Twice in a year	32	30	>0.1
Once in a year	8	8	
Absent	6	8	

Total	100	100	

PALPITATION	GROUP1 Number	GROUP2 Number	p value
In every month	28	50	
4 times a year	16	14	
Twice in a year	4	12	<0.001
Once in a year	_	6	
Absent	52	18	
Total	100	100	

ANXIETY	GROUP1 Number	GROUP2 Number	p value
Present	4	46	
Absent	96	54	<0.001
Total	100	100	

RESP. DISCOMFORT	GROUP1 Number	GROUP2 Number	p value
In every month	4	8	
4 times a year	24	16	
Twice in a year	12	14	>0.1
Once in a year	_	6	
Absent	60	56	
Total	100	100	

SKIN COLOR	GROUP1 Number	GROUP2 Number	P value
Normal	40	20	
Pale	48	60	>0.1
Yellowish	12	20	
Total	100	100	

SCLERA	GROUP1 Number	GROUP2 Number	P value
Normal	16	12	
Yellowish	84	88	>0.1
Total	100	100	

PALLOR	GROUP1 Number	GROUP2 Number	p value
Present	34	64	
Absent	66	36	<0.002
Total	100	100	

LIVER	GROUP1 Number	GROUP2 Number	p value
Non palpable non tender	80	70	
Non palpable tender	8	8	> 0.1
Palpable non tender	8	10	
Palpable tender	4	12	
Total	100	100	

SPLEEN	GROUP1 Number	GROUP2 Number	p value
Non palpable non tender	50	50	
Non palpable tender	4	6	
Palpable non tender	14	12	>0.1
Palpable tender	32	26	
Splenic atrophy	_	6	
Total	100	100	

LEG ULCER	GROUP1 Number	GROUP2 Number	p value
Present	_	4	
Absent	100	96	>0.1
Total	100	100	

CARDIOMEGALY	GROUP1 Number	GROUP2 Number	p value
Present	_	4	
Absent	100	96	>0.1
Total	100	100	

SPLENIC SEQUESTRATION SYNDROMES	GROUP1 Number	GROUP2 Number	p value
Present	32	30	
Absent	68	70	>0.1
Total	100	100	

BONE PAIN	GROUP1 Number	GROUP2 Number	p value
Present	92	100	
Absent	8	_	<0.05
Total	100	100	

BACKACHE	GROUP1 Number	GROUP2 Number	p value
Present	62	92	
Absent	38	8	<0.001
Total	100	100	

DACTYLITIS	GROUP1 Number	GROUP2 Number	p value
Present	26	38	
Absent	74	62	>0.1
Total	100	100	

PAIN IN FEMURE AND HUMERUS HEAD	GROUP1 Number	GROUP2 Number	p value
Present	40	80	
Absent	60	20	<0.001
Total	100	100	

PAINFUL CRISES	GROUP1 Number	GROUP2 Number	p value
Present	60	94	
Absent	40	6	<0.001
Total	100	10	

CHEST SYNDROME	GROUP1 Number	GROUP2 Number	P value
Present	56	68	
Absent	44	32	>0.1
Total	100	100	

PAINFUL SWELLING OF HAND	GROUP1 Number	GROUP2 Number	p value
Present	14	36	
Absent	86	64	<0.05
Total	100	100	

PNEUMOCOCCAL INFECTION	GROUP1 Number	GROUP2 Number	p value
Present	12	10	
Absent	88	90	>0.1
Total	100	100	

CHOLELITHIASIS	GROUP1 Number	GROUP2 Number	p value
Present	2	14	
Absent	98	86	<0.05
Total	100	100	

DISCUSSION

Majority of the patients in both groups suffered pain in chest, limb, joint and abdomen area. Children suffered more from abdominal pain and less joint pain, which is reverse in case of adults. Adults suffered more from joint pain. It is may be due to adults were exposed to more physical labors leading to occlusion of red blood cells in joint curves producing pain in joint. Both the groups of patients suffer evenly from fever. Though adults were exposed to more physical works the environmental factor and expose to infection is same in both groups. So affection of fever is somehow equal in both groups. Among adults 50% of patients experienced palpitation in every month where as among children 52% had no palpitation. Presence of fetal hemoglobin in children is quite helpful to fight against anemic condition, but fetal hemoglobin is lacking in adults along with rapid destruction of RBCs leading to palpitation. The skin of 60% of adult patients was pale and 20% had yellowish color, where as in children 48% had pale skin, 12% had yellowish skin and 40% had normal skin. This demonstrates the severity of the disease in adults. In both groups, majority of patients had yellowish sclera. Sickle cell anemia is a disease in which the serum bilirubin level is always high than normal. This usually gives a permanent yellowish tint to the sclera and skin. As the sclera contains collagenous tissue the pigmentation sometimes becomes permanent. So irrespective of the age group the sclera showed yellowish discoloration. In both groups 50% of patients had normal spleen. 32% of children and 26% of adults had palpable tender spleen, 6% of adult patients had splenic atrophy. Splenic atrophy or autosplenectomy shows the severity of condition in adults. In splenic sequestration syndrome there is sudden onset of pain along with splenomegaly. Both the group patients similarly affected from this syndrome. Among adults all the patients suffered from bone pain and 92% of pediatric patients had this complain. 38% of adult patients and 26% of pediatric patients had pain and swelling in fingers. Adults are more exposed to manual work, which lead to vasoocclusive processes in fingers leading to pain and swelling. This exhibits the severity in adults. 80% of adult patients had pain in femure and humerus head where as only 40% of pediatric cases described this complains. As the disease become chronic day by day the chances of a vascular necrosis is more

producing pain in the femur and humerus head. Among adults 94% of patients had painful crises and among children 60% had this complain. Adults suffered more due to vasoocclusion and destruction of RBCs leading to painful crises. Sudden occlusion of the pulmonary vessels leading to breathing difficulty and pain in chest is one of the commonest phenomena in Sickle cell anemia. In this study 68% of adult patients and 56% of pediatric patients elicited this pain. It shows the severity in adults compare to children. This study displayed equivalent pneumococcal infection both in children and adults. Both the groups of patients belong to the same locality i.e. tribes of Wayanad, exposed to same environment and the hygiene level is also similar. So pneumococcal infection affects both the groups equally. Among adults 14% of patients had cholelithiasis, whereas only 2% of pediatric patients had gall bladder stone. In adults the destruction of RBCs is more leading to deposition of bile salts and bile pigments in gall bladder later on which form stone.

CONCLUSION:

Sickle cell anemia itself is a grave disease, but the clinical severity is more in adults as they are exposed to aggravated factors more compare to children. During conducting the study, adult cases finished initially due to availability of cases but it took more time to finish the pediatric cases. Due to increase literacy level now a days normal are ready to marry sickle cell anemia patient. So offspring are either becoming normal or carrier. It is one of the best ways to reduce this disease from society.

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