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RETROSPECTIVE STUDY OF HAEMATOLOGICAL PROFILE OF SICKLE CELL DISEASE PATIENTS AT TERTIARY CARE HOSPITAL

Pathology

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ABSTRACT

INTRODUCTION: The aim of this study was to evaluate the haematological profile of patients of sickle cell disease from tertiary care hospital in western Vidarbha. This study was carried out in the Department of Pathology between the period January 2014 to December 2015.

MATERIAL & METHODS: Haematological profile of 92 patients was studied during two year period which includes 47(51.08 %) males & 45(48.91%) females. Most patients were from low socioeconomic background. Study includes the haematological parameters of the symptomatic patients attending outpatient departments of Medicine, Paediatric & Gynaecology departments. Venous blood samples were collected in ethylele diamine tetraacetic acid bulb from the patients. Haemtological parameters were evaluated using Cellenium 19 auto analyser. Samples were also tested for sickling test, solubility test, Hb electrophoresis using Hydrasys 2 gel elecrtphoresis apparatus.

RESULT: Mean age of presentation in males was 16.65 & for females it was 18.08. Most of these patients were in the age group of 6 to 20 years. Average haemoglobin percentage was 9.32 gm%, MCV was 84.37 fl, MCH was 24.59 pg, MCHC was 29.16 gm%, HbF was 17.34%.

CONCLUSION: Sickle cell disease is one of the most common severe monogenic disorder in the world. In this study it was found that, these patients had moderate to severe anaemia with high value for foetal haemoglobin. Previous studies indicate variation in haematological parameters from region to region. Thus more imphasis is needed to encourage regionwise study of haematological profile in these patients.

KEYWORDS

Haematological profile, Sickle cell disease, Anaemia.

INTRODUCTION

Sickle cell disease is one of the most common severe monogenic disorder in the world[1]. It is most common among the population whose ancestors come from sub Saharan Africa, India, Saudi Arabia & Mediterranean countries.[2,3] 50% of world population of sickle cell disease resides in India. The average frequency of sickle cell gene ranges between 22to 44%.[4]The disease is most prevalent in the low socioeconomic, tribal population particularly scheduled caste & scheduled tribes of Maharashtra. These groups reside in the remote hilly areas where health facilities are inadequate. High prevalence of sickle cell gene is also found in tribal communities of Gujrat. In Maharashtra, the sickle gene is widely found in Vidarbha region, North Maharashtra & some part of Marathwada.

Haematological profile in sickle cell disease is extremely variable& less data is available. So this study was undertaken to determine the haematological profile of sickle cell disease patients &compare the findings with other studies.

MATERIAL & METHODS:

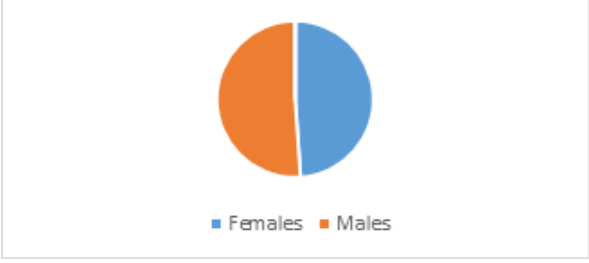
This study was carried out in the department of Pathology, Government Medical College Akola from January 2014 to December 2015. Haematological profile of 92 patients was studied during two year period which includes 47 (51.08 %) males & 45 (48.91%) females. Study includes the haematological parameters of the symptomatic patients attending outpatient departments of Medicine, Paediatric & Gynaecology departments. Patients with the history of blood transfusion within last three months were excluded from the study. Under all aseptic precautions 2-3 ml venous blood samples were collected in ethylene diamine tetraacetic acid (EDTA) bulb from the patients. Haematological parameters include Haemoglobin (Hb), Mean Corpuscular Volume(MCV), Mean Corpuscular Haemoglobin (MCH), Mean Corpuscular Haemoglobin Concentration (MCHC) were measured using Cellenium 19 auto analyser. Sickling and solubility tests were performed. Hb electrophoresis was done on hemolysate by using Hydrasys 2gel electrophoresis apparatus.

Data was recorded on a predesigned proforma &was managed on Excel spread sheet.

RESULTS:

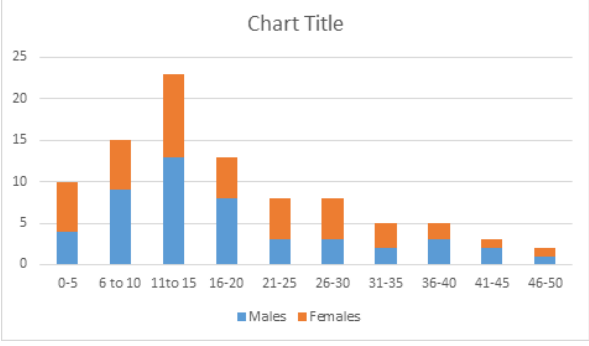
Haematological parameters of 92 patients were studied in this study which included 47 males & 45 females. **Figure 1** shows distribution of male & female patients in the study.

Figure 1. Shows male & female distribution in the study (n=92)



Maximum patients were in the age group between 11 to 15 years 9 (n=23), followed by 6 to 10 years (n=15) & 16 to 20 years (n=13). Minimum numbers of patients were observed after the age group of 30 years.

Figure 2 : Shows age wise distribution of male & female patients.



Average haemoglobin was found to be minimum in the age group 16 to 20 years (6.8 gm %), while it was maximum in age group 46 to 50 years (10.5 gm %). Average MCV was found to be minimum in the age group

31 to 35 years (77.7 fl), & higher values were observed in age group 46 to 50 years (94 fl). Lower values for MCH were found in age group 26 to 30 years(19.5 pg)& higher values were found for age group upto 5 year(30.4pg), for MCHC lower values were observed for the age group upto 5 years(26.1gm%)& higher values were found for age group 36 to 40 years (32 gm%). **Table 1** shows age wise distribution of haematological parameters.

Table 1: Shows agewise distribution of Hb, MCV, MCH& MCHC.

Sr No	Age group (years)	Hb(gm%)	MCV(fl)	MCH(pg)	MCHC (gm%)
1	0-5	7.60	79.33	30.40	26.10
2	6-10	8.35	83.63	24.05	27.00
3	11-15	8.37	82.08	21.80	27.56
4	16-20	6.85	77.88	23.14	30.50
5	21-25	7.54	91.57	24.75	27.37
6	26-30	8.70	83.66	19.50	26.80
7	31-35	8.90	77.75	24.50	32.00
8	36-40	9.15	84.40	26.20	32.80
9	40-45	8.42	89.40	23.60	31.50
10	46-50	10.50	94.00	28.00	30.00

Table 2 shows age group wise distribution of HbA2, HbF & HbSS. For HbF lower values were observed for age group 16 to 20 years (11.57%) & higher values were found for age group upto 5 years (23.83%). Lower values for sickle haemoglobin were observed for the age group 36 to 40 years (62.52%) & higher values were found for age group 6 to 10 years (76. 35%).

Table 2: Shows age groupwise distribution of HbA2, HbF & HbSS

Sr No	Age group(years)	HbA2%	HbF%	HbSS%
1	0-5	2.90	23.83	67.65
2	6-10	3.15	13.35	76.35
3	11-15	2.60	20.63	72.35
4	16-20	2.50	11.57	74.09
5	21-25	2.90	14.87	71.80
6	26-30	2.70	20.48	73.70
7	31-35	2.80	16.40	74.52
8	36-40	2.68	19.80	62.52
9	41-45	2.10	17.40	70.60
10	46-50	2.30	15.10	72.55

DISCUSSION

Haematological profile of 92 patients is presented which include 47 male & 45 female patients. The patients were from age group 6 months to 50 years. The number of female patients was more than males. Haemoglobin percentage was found to be decreased in the patients in the age group16 to 20 years. Nutritional deficiency, complications like haemolytic crisis, infections might be the possible causes of low haemoglobin level. In low socioeconomic communities like schedule caste & tribes, anaemia is commonly observed.[5,6] However steady rise in Hb percentage was observed from age group 21 to 25 years onwards. This might be due to the initiation of treatment.

Values of mean corpuscular volume(MCV), mean corpuscular haemoglobin (MCH) mean corpuscular haemoglobin concentration (MCHC) in this study are comparable with other studies.[4,7,8] In this study low values for MCHC were observed which is comparable with other studies.[9,10,11] Higher values of MCV are observed in patients of sickle cell disease. This might be because stimulation of erythropoiesis in response to haemolysis reflecting as macrocytosis. Folic acid deficiency may also cause macrocytosis[12]. Low values of MCV is seen in sickle cell disease when conditions like iron deficiency anemia or alfa thalassemia coexist[13,14]

In present study high values for HbF were observed. It correlates with the findings of other authors.[4,15] In Indian patients Arab Indian haplotype is found which is associated with high HbF values.[16]. High values of HbF in sickle cell disease result in less severe clinical presentation.[17,18]. Severity of presentation varies among sickle cell disease patients. This is due to the absence of identical pleiotropic genes. Less data is available on the phenotype of sickle cell disease therefore it is difficult to comment upon the role of HbF & complications of sickle cell disease.

CONCLUSION

It is observed in this study that the patients with sickle cell disease in this region presented with moderate to severe anaemia, low MCHC and high HbF levels. Previous studies indicate that, there is considerable variation in haematological parameters in patients with sickle cell disease. In view of availability of limited data regarding variation in haematological parameters in these patients, more extensive cohort studies are required

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