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Clinical and haematological Findings in Sudanese patients With sickle cell disease Attending the hospitals in Elobeid, Kordofan

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ABSTRACT

Objectives: This study aimed at assessing the clinical features and haematological parameters in sickler Sudanese patients attending El Obeid Hospitals in North Kordofan, Sudan.

Materials and Methods: Eighty five patients with homozygous sickle cell (SS) disease, (both sexes) were included in this study. Their age ranged between 6 months to 42 years. Forty persons were used as control.

Clinical manifestations were recorded at examination. Freshly obtained blood samples from patients and control were used to estimate haematological parameters which included: haemoglobin concentration (Hb), packed cell volume (PCV), red blood cell count (RBC), white blood cell count (WBC), mean corpuscular haemoglobin (MCH), mean corpuscular volume (MCV), and erythrocyte sedimentation rate (E.S.R).

Results: The clinical investigations revealed that all patients presented with painful crisis, 96.5% had chronic anemia, 84.7% had recurrent malaria and 81.2% had different infections (76% was chest infection). 32.9% of the sickle cell disease (SCD) patients had no blood transfusion, while the rest had blood transfused once to five times. Haematological measurement indicate that all SCD patients in this study were reported anaemic, and their Hb, PCV and RBC count were significantly lower ($P<0.001$)

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compared with the control group, while WBC count and MCV were significantly higher (P<0.001, P<0.01). In this study the prevalence of SCD in Misseria tribe (24.7%) was highest compared to other tribes.

Conclusion: This study affirmed the problem of sickle cell disease in Kordofan state, which should draw attention toward the need of further studies.

Key words: sickle cell disease, sickle cell anemia, Hematological changes, Sudan

ملخص

هدفت هذه الدراسة الى تقييم الاعراض المرضية ووضع مكونات الدم عند مرضى الانيميا المنجلية بمستشفيات مدينة الابيض بولاية شمال كردفان، بالسودان.

شملت هذه الدراسة 85 مريضا بالانيميا المنجلية المتماثلة من الرجال والنساء، تتراوح اعمارهم بين 6 شهور الى 42 عاما. 40 شخصا يمثلون مجموعة الضبط.

جمعت عينات الدم من المرضى ومجموعة الضبط لقياس مكونات الدم والتي تشمل: تركيز خضاب الدم، الكسر الحجمي للكريات الحمر، تعداد كريات الدم الحمر والبيض، المتوسط الكروي لخضاب الدم، المتوسط الكروي الحجمي، وتثقل الكريات الحمر..

اظهرت نتائج الفحص ان كل مرضى الانيميا المنجلية يعانون من الالم الحاد الناتج من ترسب الخلايا المنجلية في انحاء الجسم المختلفة، 96.5% يعانون من فقر الدم، 84.7% يعانون من الملاريا المتكررة، 81.2% يعانون من التهابات مختلفة (76% يعانون التهابات الجهاز التنفسي) 32.9% من مرضى الانيميا المنجلية لم يتم لهم عملية نقل دم، اما البقية فتتراوح عملية نقل الدم بين مرة الى خمس مرات.

اشارت قياسات مكونات الدم الى ان كل مرضى الانيميا المنجلية يعانون من مرض فقر الدم، اما تركيز كل من خضاب الدم، الكسر الحجمي للكريات الحمر وتعداد كريات الدم الحمر كان منخفضا انخفاض ذو معنى مقارنة بمجموعة الضبط، بينما تعداد كريات الدم البيض كان ارتفاعه ذو معنى مقارنة بمجموعة الضبط. ايضا انتشار مرض الانيميا المنجلية سجل ارتفاعا عند قبيلة المسيرية (24.7%) مقارنة بالقبائل الاخرى بشمال كردفان.

خلصت هذه الدراسة الى اثبات وجود الانيميا المنجلية في ولاية كردفان، وضرورة الاهتمام بهذه المشكلة الصحية بمزيد من البحث والدراسة.

INTRODUCTION

There are two main large foci of the sickle gene among the Sudanese population, the Baggara "Negro-Arab" tribe in Western Sudan and Southern Nilotics of Equatoria Province, Southern Sudan [1,2] . A survey reported a sickling rate ranging from zero to 18 percent among different tribes [3] . A survey done in Kordofan reported a prevalence of 18% in the Messieria Humur tribe [4], and up to 30.4% among Misserria tribe of

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Darfur [5] . Other results showed that Messeria which was an Afro-asiatic tribe possesses a high prevalence of sickle cell trait in Sudan [6]. Another study indicated that 84% of SCD patients were from Bagara tribe in Western Sudan, where HbS is a natural extension of the West African HbS belt [7].

Variation in the complications of sickle cell disease (SCD) was found in Sudanese patients which included: gallstones [8] , dacylitis, liver enlargement and cardiac complications, haemolytic anaemia [9, 7] , painful crisis of the vaso-occlusive type followed by fever, pneumonia and osteomyelitis [9] .

Haematological parameters in SCD patients in Sudan were studied extensively in Khartoum. Iron deficiency [10] , and haemolytic anaemia ([11, 12] were reported.

Because of the different movements of the tribes and the intermarriages, the disease is now seen in different regions in Sudan, so variation in clinical and haematological parameters are expected. This study aimed at assessing the clinical and haematological variables in sickler Sudanese attending El Obeid hospitals in North Kordofan, Sudan.

MATERIALS AND METHODS

This case control study was conducted on known sickler patients (HbSS) diagnosed by physicians depending on the clinical pictures and laboratory investigations (sickling test and electrophoresis). The study group were: eighty five sickle cell disease (SCD) patients attending El Obeid hospitals during May 2003 January 2004; forty subjects were used as control group. A questionnaire was designed to obtain information about age, sex, tribe, weight, height, education, occupation, SCD complications and family history. Venous blood samples were collected from each patient and transferred to polyethylene vials coated with an anticoagulant (EDTA). A manual haemocytometer and haematology analyzer were used for duplicate determinations of white blood cell count (WBC), red cell count (RBC) and platelets count according to standard methods. Sickling test, differential leucocyte count and RBC morphology examination were also done. Erythrocyte sedimentation rate (ESR) and packed cell volume (PCV) were determined. Mean corpuscular haemoglobin (MCH) and mean corpuscular volume (MCV) were calculated.

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Analysis of variance (one way ANOVA) was applied for comparison between different groups. P value was considered significant at a value less than 0.05.

RESULTS

The clinical manifestations of sickle cell disease (SCD) patients are presented in

Table 1. All SCD patients suffered from painful crises, while chromic anaemia, recurrent malaria and infections scored high prevalence in them .The common type of infection was chest infection (76.5%), and osteomyelitis (4.7%). Of the 85 SCD patients 57 (67.1%) had blood transfusion one to five times, while the rest had no blood transfusion. 63 (74%) SCD patients had Hb concentration between 6.1 9.0 g/dL, while 19 (22.4%) of the patients had Hb concentration less than 6 g/dL, which was significantly lower compared with the control group (Hb>10g/dL). 3 patients (3.5%) had haemoglobin concentration of 10g/dl.

PCV and RBCs count were significantly lower in SCD patients compared with control subjects, whereas white blood count (WBC) and mean corpuscular volume (MCV) were significantly higher (Table 2). Differential leucocyte count (Table 3) showed significant differences between SCD patients and control group. RBC morphology was found normal in 24.6% of SCD patients, while 57 (67.1%) of them suffered from anisopikilocytosis (Table 4).

Significant differences were found in weight and height between SCD patients and control group. The weight (kg) of the patients (n=85) was 27.3– 14.9 whereas that of the control (n=40) was 57.3– 18.5 (P<0.001).

The height (cm) of the patients was 131.4– 29.3 whereas that of the control was 156.6– 16.2 (P<0.001).

The age and sex distribution is presented in Table 5. The ratio of males to females was 1.18:1 for sickle cell patients. Most of the cases of SCD (68.3%) were in patients less than 15 years old.

The ethnic groups showed highest prevalence of SCD in Messeria tribe (24.7%), followed by Hawazma tribe (20.0%), Barno, Falata and Dagese (16.5%), Bederia (14.1%), Gawammea (10.6%), Bani Halba (7.1%), Nuba (5.9%) and Hessanat and Maganeen (1.2%).

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Table 1. Clinical findings and complications in the studied sickle cell disease patients

Clinical findings and complications	(n=85)	%
Painful crisis	85	100.0
Chronic anaemia	85	100.0
Recurrent malaria	72	84.7
Infections	69	81.2
Foot hand syndrome	56	65.9
Jaundice	47	55.3
Ocular problem	23	27.1
Haematouria	12	14.1
Leg ulcer	12	14.1
Priapism	13	15.3
Gallstones	5	5.9

n= Number of Patients

Table 2. Haematological parameters in patients and control group (Mean–SD)

Variable	Patients (n = 85)	Control (n = 40)	Significance
Hb (g/dL)	7.12 – 1.3	12.82 – 1.33	S ***
P.C.V (%)	21.7 – 4.2	38.9 – 4.4	S ***
RBC count (10^{12}/L)	2.51 – 0.78	4.74 – 0.46	S ***
MCV (fl)	92.5–31.81	80.1–3.7	S **
MCH (pg)	30.76 – 10.05	29.3–1.24	N.S
E.S.R (mm/1 hr)	21.2–20.6	17.6–6.6	N.S
WBC count (10^9/L)	12.57–4.56	5.88–1.49	S ***

n= Number of subjects
* = Statistically significant P < 0.05
** = statistically significant P < 0.01
*** = statistically significant P < 0.001
N.S = statistically not significant SD = standard deviation

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Table 3. Differential leucocyte count in the patients and control (Mean – SD)

Variable	Patients (n = 85)	Control (n = 40)	Significance
Neutrophil	50.34–5.1	56.1–5.19	S ***
Lymphocyte	43.85–4.95	38.9–5.31	S ***
Monocyte	3.16–1.55	4.06–1.43	S ***
Eosinophil	1.35–1.55	0.87–0.75	S ***

n= Number of subjects
* = Statistically significant P < 0.05
** = statistically significant P < 0.01
*** = statistically significant P < 0.001

Table 4: Red blood cells morphology in sickle cell patients and control groups n= Number of subjects

Finding	Patients (n=85)		Control (n=40)	
	N	%	N	%
Normocytic normochromic	21	24.6	40	100
Anisopoikilocytosis	57	67.1	0	0
Normocytic hypochromic	3	3.5	0	0
Microcytic hypochromic	2	2.4	0	0

Table 5. Age and sex distribution of the studied SCD patients

Age group (year)	Male		Female		Total	
	No	%	No	%	No	%
<2	2	4.3	3	7.7	5	5.9
2-8	22	47.8	14	35.9	36	42.4
9-15	9	19.6	8	20.5	17	20.0
16-22	4	8.7	4	10.3	8	9.4
23-29	3	6.5	6	15.4	9	10.6
> 29	6	13.0	4	10.3	10	11.8
Total	46		39		85	

DISCUSSION

In this study all patients were homozygous sickle cell anaemia patients (HbSS).

The common clinical findings observed were chronic anaemia, and painful crisis.

These two findings were common in SCD patients [12-16], but not common in Indian patients [17]. Recurrent malaria was also reported in this study, which indicates that the occurrence of malaria (P. Falciparum) in homozygous sickle cell (SS disease) is almost certainly a major determinant of morbidity and mortality [18] ,in contrast to the (AS) trait

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who have resistance to this parasite. Chest infection was reported high in this study. This agrees with Al Dabbous, 2002; Stuart, 1999 [19, 20] but not in line with other studies that reported various infectious diseases other than chest infection [7,12] . Due to excessive haemolysis of sickled RBCs and according to the WHO criteria (Hb<11.012.0 g/dL), all SCD patients in this study were anaemic, and their mean Haemoglobin

(Hb) concentration was significantly (P<0.001) lower compared with the control group. This result agrees with Bayoumi, 1988; Awad, 1992; Nduka, 1993; Kar et al, 1986 [7, 21, 22] . Most of the patients in this study had blood transfusion once to five times. This may explain the presence of normocytic normocromic erythrocytes in SCD patients. Despite the highly significant difference in mean corpuscular volume (MCV) between the two groups; it falls within the reference range. This finding agrees with Beharman (1996) [23] , who also found that RBC morphology in SCD patients was normochromic normocytic, and their platelets were increased, while in this study the anisopoikilocytosis was reported high, and only 27 (31.8%) SCD patients had increased platelets. White blood cell count (WBC) was significantly higher in SCD patients. Our finding that packed cell volume (PCV) and red blood cell count (RBC) were significantly lower in SCD patients compared with control subjects is in line with ElHazmi et al, 1987; Nduka, 1993 and Kar et al, 1986 [17,22,24] . In general, erythrocyte sedimentation rates (E.S.R) in SCD patients is low probably because of abnormal shape of sickle cells that prevent relax formation. In this study E.S.R was higher in SCD patients compared with control. A significant difference was found in weight and height between SCD patients and control group. This result agrees with others who found that impaired growth is common, and there is early decrease in height and weight [25].

The prevalence of SCD was found higher in Messeria which was an Afro-asiatic tribe followed by Hawazma. This result is in agreement with Atalla et al, 2003; Ibrahim, 1970; and Vella, 1964, [4-6] while another study found that the prevalence of SCD in Bagara tribe was higher compared with other tribes [7] . This difference can perhaps be accounted for by the small sample size in these studies which may not be representative of the tribes of the whole population.

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