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Original Article

Assessing clinical laboratory funding of sickle cell disease and other associated disorders in Khuzestan province

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Abstract

Objective: The aim of this study was to assess clinical laboratory funding for differential diagnosis of sickle cell disease (SCD) and other associated disorders for better understanding of clinical types and prevention of sickling events.

Patients and Methods: This is a descriptive crossed-sectional study that analyzed the peripheral blood film, sickle cell preparation, hemoglobin electrophoresis, CBC, RBC indices serum ferritin related to clinical features on referral cases and their family from different parts of Khuzestan province. Twenty-eight cases (SS) and others associated disorders, 16 (SIDA), sickle – thalassemia, 32 (Sβ$^+$), 30 cases of (Sβ °) and 31(AS), were studied from 1999 to 2002.

Results: Family reviewed showed 97% of sickle carrier states were among the ethnics Arabian origin, 10% Arab – Persian half breeds only 3% from old Persia in origin. Elevated HbF was seen in most of the patients who were prominent in SS rather than other associated heterozygote and sickle traits. About 85% of patient had HbF level more than 13%. MCV>80 MCH>27 were obtained in SS and sickle traits. Baseline WBC counts (neutrophils polymorphoneuclear) in SS individuals are elevated.

Conclusions: General appearance of these patients usually good looking Caucasian boys and girls with a diagnosis of SCD others associated disorders.

Key words: Laboratory funding; Sickle Cell Disease; Thalassemia; HbF

Introduction

Clinical practice shows that hereditary sickle cell disease (SS) and others associated disorders such as sickle –Thalasseia (S β$^+$, S β°), sickle/ Iron deficiency anemia (SIDA), Sickle trait (AS) are the most common congenital hemoglobinopathies found by medical practice in the south west region of Khuzestan and neighboring regions[1]. Most of SCD is often mild with a high level of HbF1. Furthermore, there is a lack of sufficient published research works and data on the nature, prevalence and assess of clinical laboratory pattern, hematological feature and distinct mutation of SCD in South west Iran, where in medical practice the frequency of SCD associated disease is seems to be high[2-6].

The aim of this study was to assess clinical laboratory funding for differential diagnosis of SCD and other associated disorders in our medical practice in this region for better understanding of clinical types and prevention of sickling events, so we recorded serials of clinical laboratory funding related to their clinical manifestations as follows.

Patients and Methods

To approach this hypothesis serial evaluation of patients and their family were carried out as a descriptive crossed-sectional study. Analysis of blood smears and quick sickle cell preparations by 10% sodium bisulfate were performed in one hour and then fallowed for 24 hours. Hemoglobin electrophorus analysis was done by Helena apparatus. CBC, RBC indices by Helena electronic cell counter and clinical features conducted SCD and others concurrent associated disorders, were done on 28 cases (SS), 16 (SIDA), 32 (S β $^+$), 30 (Sβ °) and 31(A S), from 1999 to 2002. Among the referral cases and their family to Ahvaz

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Thalassemia–sickle cell center Shafa Hospital affiliated to Jundishapur University of medical sciences[2-4]. First of all an oral discussion was done to the patients and their family then an informed written consent was obtained from each participate and then samples of blood was taken.

Statistical analysis

All data were presented as mean and Standard deviation. The data were analyzed using SPSS 13.0.

Results

Compression characteristic clinical laboratory data finding of 28 (SS) cases and others 139 concurrent associated diseases (SB^+, SB°) are on the figures[1-5]. Geographical distribution showed that the numbers of patients were on an increasing trend from the southwest and south toward the center and then continued to the eastern part of Khuzestan province, these regions are the areas where most ethnic Arabian people mainly are living there. Family SCA screening showed 97% of sickle carrier states are among the ethnics Arabian origin, 10% Arab–Persian half breeds, 3% of older ethnics Persian origin (Figure1) the elevation is more prominent in SS rather than other associated heterozygote and sickle traits. About 85% of patient had HbF level more than 13% higher level. HbF is a benign factor of SCD and other associated disorders clinical symptom and prognosis. Elevated HbF is seen in most of the patients, the elevation is more prominent in SCD rather than other heterozygote and associated sickle traits (Figure 3). CBC and red cell indices were done by the electronic cell counter, MCV>80100, are obtained in the SCA and sickle traits, otherwise MCV<80-60, in the SB°, SB^+, SCD+ Iron deficiency (Figures 2 and Table 1). White Cell counts were almost elevated in patients with sickle crisis they were admitted to the hospital in an abdominal emergency state, hand-foot syndrome, hemolytic crisis, hypoxia, dehydration, and chest syndrome (Figure 4).

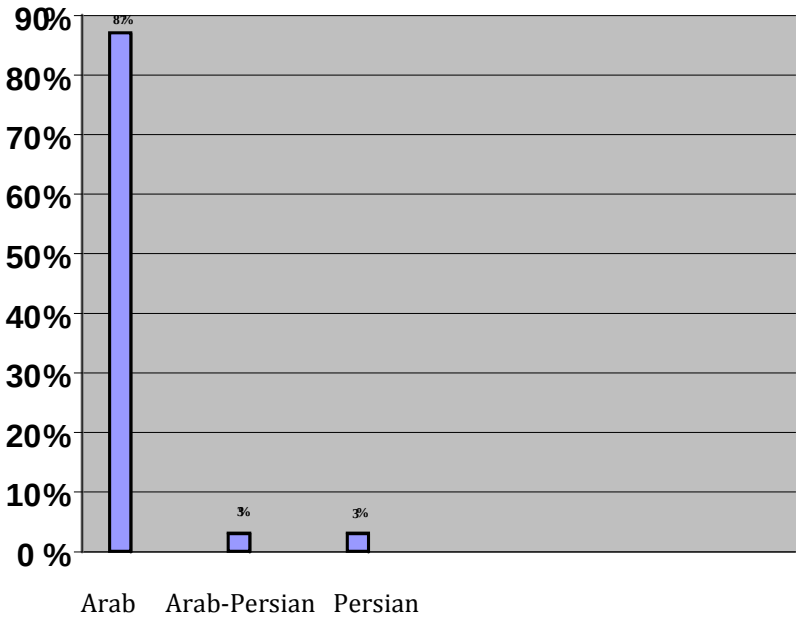

Figure 1. Arab, Arab-Persian and Persian distributions of sickle cell in Khuzestan

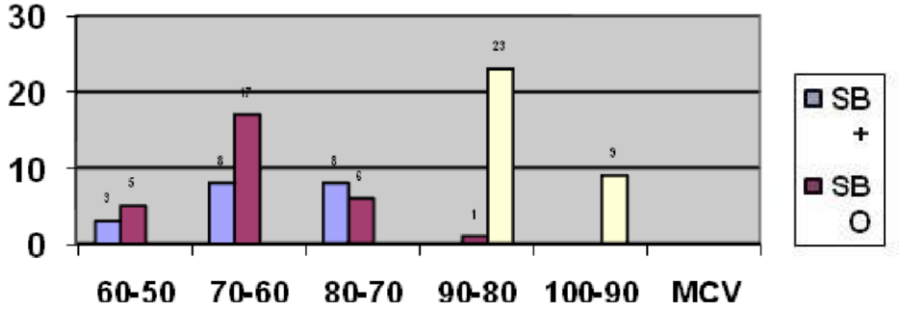

Figure 2. Variable MCV in the SCD and associated disease

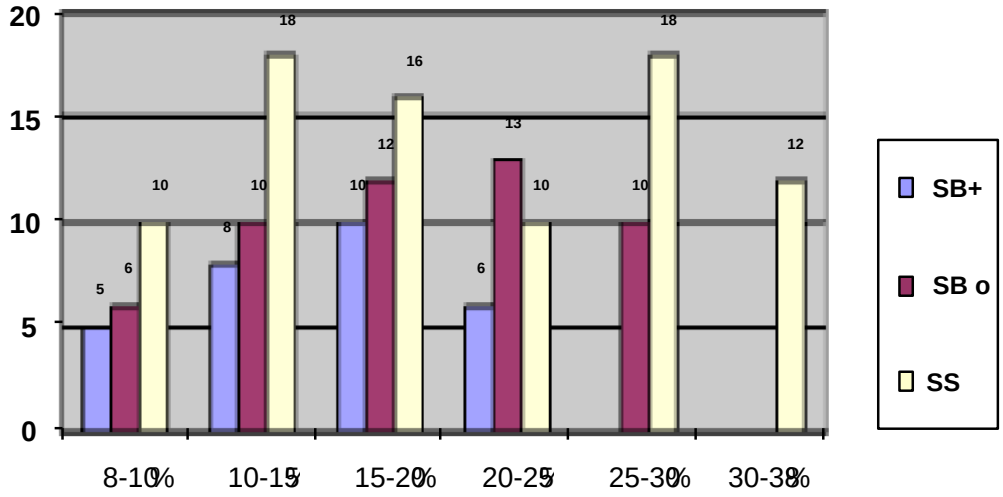

Figure 3.Variable F .Hemoglobin in the SCD and associated diseases

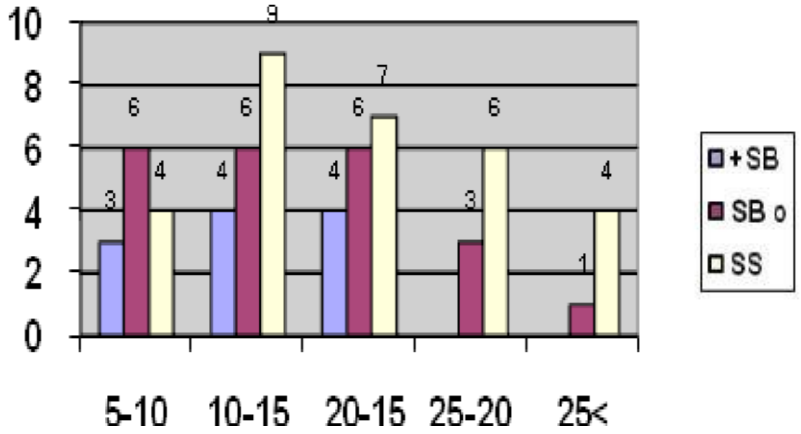

Figure 4. Patterns of leukocytosis in SCD and associated disease according to 1000

Table1. Hematological indices for patients and control groups

Groups	No.	Hbgr/dl	RBCx10^{12}/l	MCV(fl)	MCH (pg)	MCHC(g/dl)	HB-electrophoresis
SS	28	8.65±3.8	3.32±0.51	89.2 ±6.51	29±2.62	33.2±2.25	S,F.A_2.
SB^0	32	8.57±3.4	4.20±0.63	63.5±4.21	20±3.6	33.2±1.6	S,F.A_2
SB+	30	8.86±2.7	4.76±0.43	64.4±3.92	21±2.18	32.3±3.4	S,F,A_1,A_2
SA	51	13.21± 1.23	5.16±3,23	86.2±3.64	28.6±2.2	32 .8±1.8	S.A1.A2,F.
SS+ Iron Deficient	16	8.17±2.4	4.10±0.43	62.4±23.12	20±1.6	31.21±1.3	S,F.A_2 .

Discussion

In this study clinical finding showed that Khuzestan or Iran SCD is to be different from the Black Africans negro. The general appearance of the patients usually good looking Caucasian boys or girls with a diagnosis of SCD and other concurrent associated sickle Thalassemia (SB^+, SB°)[7]. The study showed 94% of sickle carrier state is among the ethnic Arab tribes with high rate consanguineous marriage (Figure 1). Sickle trait usually without clinical manifestation and with normal RBC indices, Without hemoglobin electrophorus nobody can differentiate Sickle trait from normal variations, so sickle trait might skipped through pre - martial Thalassemia health screening program[2]. Because there is a long list complication of pregnancy in SCD[8], and in double compound heterozygote of (S/ β Thal) disease[9], to prevent complications of SCD other sickle associated disorders our estimation showed official obligatory Hb electrophorus should be administered to Iran Thalassemia health pre-martial Thalassemia health screening program in high prevalence Sickle cell regions, Khuzestan Province and others south of Iran at the north border of Persian golf and Oman sea[1-3,8-9] . It is known that CSD does not usually manifest clinically in babies born until the age of about six months, when the level of HbF in blood has reduced considerably. Therefore, a high proportion of HbF in blood as an ameliorating factor to makes lower risk of sickling. The last most important variable affecting the clinical severity of SCD is due to the level of HbF[10]. HbF is heterogeneously distributed among red cells SS individual. HbF does not crystallize like Hb SS in the presence of hypoxia to causes sickling red cells. Because HbF is a potential inhibitor of polymerization, F cells are resistant to sickling and survive much longer in circulation than non-F cells[10]. The ability to produce HbF varies considerably among different population. Therefore, among populations in the eastern region of the Saudi –Arabia and India, SS patients have high HbF levels and relatively mild disease, high level. HbF is a benign factor of SCD and other associated disorders in clinical symptom and prognosis[9-10]. These epidemiological observations have been a powerfull incentive for the development of pharmacological agents for induction of γ-globin expression HbF production[11]. HbF values can also be valueable in the monitoring of hydroxyurea therapy, which raises the HbF level[12].

There has been increasing evidence that circulatory WBCS Particularly neutrophil play a role in SCD disease Baseline WBC counts in SS individuals are elevated neutrophil, polymorphoneuclear. Activation of neutrophils is seen in the steady states and during vasoocclusive crisis. The degree of elevation is directly related to early mortality and increased frequency of acute chest syndrome and is a predictor of clinical severity in infants[12]. Moreover, neutrophil showed enhanced adhesion to endothelium may reduced blood flow and in association with sickled erythrocytes can cause microcirculation and crisis It is now recommended that all newborn at risk to be screened for SCD[13].

Conclusion

Analysis of blood smear, automated hematological results, RBC indices, Hb electrophoresis, sickle cells preparation are simple ways for diagnosis of SCD and other differential concurrent associated diseases[14].

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