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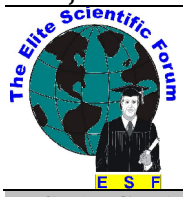

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RESEARCH ARTICLE

Effect of β-Thalassemia on Hematological and Biochemical Profiles of Female Patients

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ABSTRACT

Thalassemia is one of the most common monogenic disorders all around the world. The disease alters the hematological and biochemical parameters of the patients important in future care of the patient for plan of blood transfusion, iron chelation therapy and other life saving strategies. The study describes socio-economic, hematological and biochemical profiles of female thalassemic patients. Fifty female thalassemic patients were selected from different thalassemic centers in Punjab (Pakistan). The patients had mean age 14.6±3.09 years and body weight 35.7±10.4 kg. Siblings of 82% patients had at least one type of thalassemia as the consanguinity was 62%. Most of the patients belonged to lower class 58%, spleenectomy 60% and delay in puberty and secondary sex characters 82%. The mean±SD values of CBCs were: WBCs $35\times10^3 \pm 10\times10^3/\mu L$, neutrophils 32 ± 14.8%, hematocrit 28 ± 4%, MCHC 31±5 g/dL, RBCs $3\times10^6\pm0.6\times10^6/\mu L$, Hb 8±1 g/dL, platelet $322\times10^3 \pm 65\times10^3/\mu L$, RDW 43% ± 11% and ESR 50±11 mm/h. The biochemical parameters were: alanine transaminase 119±10 U/L, aspartate transaminase 77±23 U/L, alkaline phosphatase 366±150 U/L, triglycerides 108±38 mg/dL, cholesterol 104±35 mg/dL, total protein 5.4±2.1 g/dL, albumin 4.5±1 g/dL, serum creatinine 3.2±1 mg/dL, urea 3.5±1.3 mg/dL, total bilirubin 2.4±1.3 mg/dL and serum ferritin 3255±1594 µg/L. Both CBC and biochemical parameters of the patients significantly deviated from the normal values ($P < 0.05$). The altered hematological and biochemical parameters may change iron chelation response in the patients. Therefore, CBC and biochemical parameters should be considered for future plan of transfusion and iron chelation therapy.

INTRODUCTION

Thalassemia is one of the most common monogenic disorders that affect synthesis of globin chain in the patients. The disease is more prevalent in Mediterranean countries and Southeast Asia. The large population size, consanguineous marriages and high birth rate in Pakistan make the disease highly prevalent. Thalassemic homozygotes born in Pakistan are approximately 7000 each year with an overall carrier frequency of 5-5.65 % (Arif et al., 2008; Baig et al., 2006). Thalassemic patients produce inadequate amount of hemoglobin due to reduced production or absence of β globulin chain. The resulting excess of α chain precipitates in erythroid precursors and lead to premature death (Danjou et al., 2011; Galanello and Origa, 2010; Pan et al., 2007). Almost all types of thalassemia can be diagnosed by hemoglobin level and other hematologic analyses.

The management of thalassemia is one of the urgent concerns for the patients and has achieved great development in the world. Regular blood transfusion is one of the conventional treatments of thalassemia to keep the hemoglobin levels close to normal (Khan et al., 2010). Patients with β-thalassemia major require regular red blood cell (RBC) transfusions to support normal growth and development and reduce extra-medullary haematopoiesis (Jamuar et al., 2011).

The degradation of red blood cells and transfusion result in excessive iron deposition in body tissues which

leads to many pathophysiological conditions like expanded plasma volume, cardiac output and reduced glucose tolerance (Abdelrazik, 2007; Borgna-Pignatti et al., 2004; Cunningham et al., 2004). Thalassemia and blood transfusion affect the blood biochemistry and hematology of the patients. In this paper, we report the hematological and biochemical studies of local female thalassemic patients helpful for future plan of blood transfusion and management of the complications of disease.

MATERIALS AND METHODS

Female β-thalassemia major patients (n=50) above 12 year age were selected from Red Crescent Thalassemic Centre Faisalabad, Sundas Foundation Faisalabad, Fatimid Foundation and General Children Hospital, Multan. The sampling was carried out according to the ethical principles of Helsinki (WHO, 2008). Each patient was interviewed along with parents and mothers were preferred as respondents rather than fathers to fill the questionnaire. The research work was conducted at Department of Chemistry and Biochemistry, University of Agriculture, Faisalabad. All the patients were physically examined for body weight, age, blood pressure and body temperature before the start of study. Three mL venous blood was collected from each of the patients and divided in three parts: (a) complete blood count through hematology analyzer Sysmex Kx-21, (Germany) (b) erythrocyte sedimentation rate (ESR) and (c) biochemical parameters by clinical chemistry analyzer, Micro lab 200 (Merck, Germany).

Statistical analysis

The data was statistically analyzed through SPSS package (SPSS, Version 19). The descriptive analyses are presented in terms of means and standard deviations with range. Pearson correlation and multivariate ANOVA was performed. Differences with $P<0.05$ were considered significant.

RESULTS AND DISCUSSION

The patients belonged to different localities of Punjab like Multan, Faisalabad and Toba Tek Singh. Most of the patients belonged to rural areas and had low family income (Table 1). The mean age of β-thalassemic patients was 14.6±3.1 years body weight 35.7 ± 10.4 kg. All the patients had normal systolic and diastolic blood pressure. The patients showed physical symptoms of thalassemia and fever was prevalent in all of them.

The hematological parameters of thalassemic patients showed much variation than the reference values (Table 2). The patients were deprived of red blood cells due to their early degradation as a result of abnormal globin molecule. Most of the patients had moderate to severe anemia. This results in low hemoglobin level (8 ± 1 g/dL). The poor oxygen supply to the body tissues leads to retarded growth and other pathophysiological conditions. Pootrakul et al., 2004 reported the hemoglobin level of beta-thalassemia/HbE untreated patients as 6.6±11 g/dL. Zimmermann et al., 2008 showed Hb level 4.5-8.8 g/dL of beta-thalassemic women. The results of present study are also in comparison to mean hemoglobin level of thalassemic patients 8.4 g/dL and 8.8 g/dL (Voskaridou et al., 2011; Khan et al., 2010).

Table 1: Socio-demographic characteristics of β-thalassemia major patients

Parameters	Number of patients (n=50)
Residence	
Rural	34
Urban	16
Faisalabad	15
Multan	28
T.T. Singh	07
Socioeconomic status	
Upper	05
Middle	16
Lower	29
Education	
Literate	38
Illiterate	12
Age at diagnosis (<6 month)	31
Awareness about disease	41
Behavioral problems	12
Physical symptoms	
Pain	43
Fever	08
Pigmented skin	02
Heart complications	05
Gums	04
Disease in blood relation	41
Consanguinity	31
Blood group	
A+	02
A-	02
B+	21
B-	04
AB+	02
AB-	00
O+	08
O-	11
Transfusion	
Twice a month	16
Thrice a month	34
Spleenoctomy	30
Delay in puberty	41

The amount of white blood cells was higher than normal people. This is evident from hyper-activity of immune system of the patients receiving blood from different donors.

Table 2: Complete blood count of thalassemic patients

Parameters	Unit	Patients (n=50)		
		Mean ± SD	Min.	Max.
WBCs	(10^3/µL)	35±10	18.5	67
NEUT	(%)	31±5	14	47.2
HCT	(%)	28±4	21	39.4
MCHC	(g/dL)	31±5	21	40
MCH	(pg)	31±5	21	41
RBCs	(10^6/µL)	3±0.6	2	4.5
Hb	(g/dL)	8±1	6	9.5
MCV	(fL)	78±9	47.4	90.3
RDW	(%)	43±11	23	66
Platelets	(10^3/µL)	322±65	211	471
ESR	(mm/h)	50±11	32	69

WBCs: white blood cells; NEUT: neutrophils; HCT: hematocrit; MCHC: mean corpuscular hemoglobin concentration; MCH: Mean corpuscular hemoglobin; RBCs: red blood cells; Hb: hemoglobin; MCV: Mean cell volume; RDW: Red blood cell distributed width and ESR: Erythrocyte sedimentation rate; n: Number of thalassemic patients; Min: Minimum and Max: Maximum.

Table 3: Biochemical parameters of male and female thalassemic patients

Parameters	Unit	Patients (n=50)		
		Mean ± SD	Min.	Max.
ALT	(U/L)	119±10	30	567
AST	(U/L)	77±23	34	131
ALP	(U/L)	366±150	102	775
Glucose	(mg/dL)	112.3±9	100	120
TG	(mg/dL)	108±38	80	198
Cholesterol	(mg/dL)	104±35	83	198
Total protein	(g/dL)	5.4±2.1	2	9
Albumin	(g/dL)	4.5±1	2.1	5.6
Creatinine	(mg/dL)	3.2±1	1	8
Urea	(g/dL)	3.5±1.3	1	6
TB	(mg/dL)	2.4±1.3	1	5
Ferritin	(µg/L)	3255±1594	980	9200

ALT: Alanine transaminase, AST: Aspartate transaminase, ALP: Alkaline phosphatase, TG: Triglycerides and TB: Total bilirubin.

Rathod et al. (2007) showed that β-thalassemia is associated with mild anemia, reduced mean corpuscular volume, mean corpuscular hemoglobin (MCH) and elevated hemoglobin A2 levels. The elevated level of ALT and AST in the patients (119±10 U/L) is due to hepatic and myocardial toxicity of iron overload, a characteristic of thalassemia (Table 3). The increased ALT level is also due to prevalence of hepatitis C virus in thalassemic patients as all the patients were HCV positive. Serum ferritin as an index of iron overload was significantly high 3255±1594 µg/L than normal ($P<0.001$). The findings of this study are comparable with HCV +ve thalassemic patients which showed highly significant serum ferritin levels 4349 µg/L than 3338 µg/L in HCV -ve thalassemic patients (Khan et al., 2010). There was significant correlation among biochemical parameters like ALT with alkaline phosphatase, AST with cholesterol, creatinine with total protein, cholesterol, and total protein with glucose and cholesterol ($P<0.05$).

It has been concluded that thalassemic patients presented multiple abnormalities due to repeated transfusions, iron overload, hepatitis, cardiac and renal dysfunction. The altered hematological and biochemical parameters are important for blood transfusion and designing dose regimen for iron chelation and management of disease. Therefore, CBC and biochemical parameters should be considered for future plan of transfusion and iron chelation therapy. The study will be helpful to increase life expectancy of thalassemic patients.

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