Iraq JMS

Published by Al-Nahrain College of Medicine
ISSN 1681-6579
Email: Iraqi_jms_alnahrain@yahoo.com
http://www.colmed-alnahrain.edu.iq/

Some Observations on the Occurrence of β-Thalassemia in Mosul

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Abstract

Background β-thalassemia is found in Mediterranean, Saudia Arabia, Jordan, Egypt and Yemen. Survey in Iraq showed that β-thalassemia trait is carried by 4.5-5% of the population.

Objective To determine factors that is associated with thalassemia in Mosul district.

Methods 105 thalassemia blood transfusion dependent children with the age of 2.5–18 years attending Ibn Al-Atheer Teaching Hospital in Mosul city during 2005 were included in this study, 45 healthy subjects served as control. Blood groups, Hb, and PCV were evaluated. SI, TIBC, and TS levels were also determined.

Results The occurrence of homozygous β-thalassemia is significantly higher in the offspring of first cousin marriages."O" blood group represents a highest percentage (42.8%) among the thalassemic patients. Hb, PCV and TIBC were below the control measurements. Whereas SI and TS were above that of the controls.

Conclusions Consanguinity seems to be increasing the size of the disease due to the limited health education of the parents about the disease.

Key words Thalassemia, Hb, PCV, SI, TIBC, TS

Introduction

Thalassemia has been classified by the world health organization as a major public health problem [(1)]. It occurs throughout the world and regarded as one of the major health problems in endemic regions as the Mediterranean countries, Middle East, North Africa and Asia [(2-4)]. The thalassemia trait is characterized by a reduction in or absence of synthesis of one or more globin chains in the hemoglobin (Hb) molecule [(5)]. B-thalassemia is found in Arabic countries especially those which are located on the Mediterranean, Saudia Arabia, Jordan, Syria and Yemen [(6)]. Survey in Iraq showed that β-thalassemia trait is carried by 4.5-5% of the population [(7)]. The disease remains incurable with complications result from iron overload as result of blood transfusion and increased intestinal absorption of iron[(8)].

The aim of the present study is to determine some factors that are associated with thalassemia in Mosul district such as age and gender distribution of the disease, the relation between thalassemia and consanguinity of the parents, to measure Hb and packed cell volume (PCV) before blood transfusion and to investigate the serum levels of serum iron (SI), total iron binding capacity (TIBC) and transferrin saturation (TS).

Methods

One hundred and five thalassemia blood transfusion dependent children with the age of 2.5-18 years (62 males and 43 females) attending Ibn Al-Atheer Teaching Hospital in Mosul city during 2005 were included in this study. The local ethical committee approved the study. Fifty four healthy children with the

age of 4-17 years served as a control group. Five mls of venous blood were drawn from the cubital vein using disposable needles and syringes without using tourniquet. Blood groups were determined according to the methods of Rowley and Milkins [9]. PCV and Hb were estimated using the methods of Bain et al, [10]. SI and TIBC conducted according to Varely et al, [11].The mean, standard deviation, correlation coefficient and t-test were used for statistical analysis. The differences were considered significant when $p \leq 0.05$.

Results

Age and gender distribution of patients with homozygous β-thalassemia is shown in Table 1. A mean age of 8.91 years was recorded with a significantly higher ($p < 0.001$) incidence among males. The effect of consanguinity on the occurrence of homozygous β-thalassemia is shown in Table 2. The percent of thalassemia is significantly higher ($p < 0.001$) in the offspring of first and second cousin marriages in comparison with unrelated marriages.

As concerning the hematological parameters, Table 3 shows the blood group distribution in homozygous β-thalassemia. O^+ blood group was the highest (42.86%) among the thalassemic patients.

Hb and PCV levels were significantly ($p < 0.05$ and $p < 0.001$) below normal respectively in all of the 105 thalassemic patients Table 4. The mean Hb value was 8.04±1.1 g/dl while the mean PCV % was 25.6±3.5 in comparison with 13.5±0.9 and 39.7±2.8 in the control group respectively.

Referring to the iron status, Table 5 shows the iron levels in thalassemic patients and control groups. In the thalassemic patients, the mean SI was 183.4±44.3 µg/dl, the mean TS was 79.1±21.4 and the mean TIBC was 239.2±46.2 µg/dl, in comparison with 101.5±21.1 µg/dl, 32.7± 8.1 and 302 ± 45.1 µg/dl in the control group respectively. The mean SI and TS in the thalassemic patients were significantly ($p < 0.001$) higher than in the control group; whereas the meanTIBC was significantly ($p < 0.001$) lower than in the controls.

Table 1. Age and gender distribution of patients with homozygous β-thalassemia

Gender		Age(year)				Total
		< 2-6	> 6-10	> 10-14	> 14-18	
Male	No.	22	16	19	5	62
	%	20.9	15.2	18.1	4.7	59.1*
Female	No.	13	11	10	9	43
	%	12.3	10.4	9.5	8.5	40.9
Total	No.	35	27	29	14	105
	%	33.3	25.7	27.6	13.3	100

*$p < 0.001$ in comparison with female.

Table 2. Consanguinity and β-thalassemia major

Consanguinity	No.	%
Offspring of first cousin marriage	58	55.2
Offspring of second cousin marriage	17	16.2
Offspring of far relative marriage	13	12.4
Offspring of unrelated marriage	17	16.2
Total	105	100

Table 3. Blood group distribution in homozygous β-thalassemia

Blood group	Patient		Mosul Study [20] n=3177	Iraq Study n=24063
	No.	%	%	%
A^+	28	26.67	28.8	30.2
B^+	18	17.14*	23.9	26.7
O^+	45	42.86*	34.7	34.7
AB^+	9	8.57	12.6	8.4
A^-	3	2.86		
B^-	1	0.95		
O^-	1	0.95		
Total	105	100	100	100

**p* < 0.05 in comparison with Mosul and Iraq study

Table 4. Hematological data in patients and control group according to gender

Group	Gender	No.	Hb (g/dl)	PCV%
			Mean ±SD	Mean ±SD
Patients	M	62	8.2±1.05	26.1±3.2
	F	43	7.6±1.2	24.9±4.8
	Total	105	8.04±1.1*	25.6±3.5**
Control	M	28	13.9±0.8	40.6±2.6
	F	24	12.9±0.8	38.7±2.6
	Total	52	13.5±0.9	39.7±2.8

**p* < 0.05 in comparison with the control, **p* < 0.01 in comparison with the control.

Table 5. Iron levels in homozygous β- thalassaemia and normal subjects

Group	Gender	SI (µg/dl)	TIBC (µg/dl)	TS (%)
		Mean ±SD	Mean ±SD	Mean ±SD
Patients	M	192.1±45.8	237.4±44.80	82.9±20.04
	F	169.8±38.4	241±48.8	73.2±22.5
	Total	183.4±44.3*	239.2±46.2*	79.1±21.4*
Control	M	113.4±21.4	296.8±48.3	37.7±9.1
	F	91.8±20.8	310.1±42.1	29.6±7.1
	Total	101.5±21.2	302±45.1	32.7±8.1

**p* < 0.01 in comparison with the control.

Discussion

β-thalassemia is a common genetic disorder in the Mediterranean countries and the Middle East [2,12]. The present study focused on the occurrence of this disease in "Mosul" due to the appreciable number of patients with thalassemia major encountered in Mosul pediatric hospitals all around the year, the known endemicity of malaria in the area to which the prevalence of the disease probably related since there is a possibility of an association between beta thalassemia and malaria [13] and the high rate of consanguineous marriage among the rural.

A mean age of 8.91 years of the patients was found to be on regular blood transfusion which is higher than that found in other study "7.71 years" [6]. AS concerning the gender distribution of the disease, the study showed a higher

occurrence among the males (59.04%) than in the females (40.96%) which is comparable to previous studies [6,14,15].

Consanguinity seems to play an essential role in increasing the size of the problem in Mosul since 71.4% of the patients studied were the product of marriages between first and second cousins. Al-Haj (15) found a higher results (88%) whereas a lower result (41.6%) was reported by Awad[6].

Hematological tests showed low Hb and PCV levels with a means of 8.04 g/dl and 25.6% respectively compared with 13.5 g/dl and 39.7% in the control group. These results of low Hb among patients can be explained by the limited health education of the parents about the disease ,so that , blood transfusion was used only when the patient showed clinical symptoms caused by severe anemia or simply just to sustain life [16-17] . Whereas reports from other countries focused on a supertransfusion program (maintaining Hb level above 12 g/dl) or hypertransfusion program (where the Hb level never allowed dropping below 9 g/dl) [18-19].

As compared to normal blood group distribution in both Mosul [20] and Iraqi [21] population, the present study showed that the frequency of patients with blood group "O" is higher than in the normal population; on the other hand, the frequency of patients with blood group "B" is lower than the control. No significant difference was found in blood group "A" and "AB". This result is comparable to that reported by Al-Haj Ahmed [14], but disagrees with the study conducted by Awad [6] on Mosul district.

Generalized iron loading of the organs has been a recognized complication of thalassemia major [8]. SI and TS values were found to be higher (183.4 µg/dl, and 79.1%) than in the control group (101.5 µg/dl and 32.7%). These findings are in agreement with other studies [22-24] which refer to an increase in SI and TS in addition to other parameters.In patients with thalassemia, the excess iron results not only from blood transfusion but also from increased iron absorption secondary to the ineffective erythropoiesis, which is associated with an increased plasma iron turnover [25-26].

It is concluded from this study that β-thalassemia patients were prevalent in Mosul city compared with other regions of Iraq, consanguinity is clearly obvious especially among first cousin marriages and that all patients have shown anemia which is more common in females than males.

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Received: 21st Oct. 2009, Accepted: 8th Dec. 2010.