PLASMA LEVEL OF SOLUBLE GLYCOPROTEIN IIB/IIIA (GP IIB/IIIA) IN B - THALASSEMIA PATIENTS.

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Introduction

βeta - Thalassemia is a worldwide single gene autosomal recessive genetic disorder characterized by excess unpaired alpha globulin chains and deficient or absent beta globulin chains. Excessive increase of unpaired α-globin chains in red blood cells causes hemolysis and ineffective erythropoiesis leading to chronic anemia and hypoxia, that triggers most of the symptoms that the patients exhibit.

Recently patients with thalassemia disease are described according to their transfusion requirements into transfusion-dependent thalassemia andnon-transfusion-dependent thalassemia. Thromboembolic events (TEE) are a known complication observed in thalassemic patients. Multifactorial mechanisms are tangled in the pathogenesis of TEE in thalassemia, including the combination of the classical components of the hemostatic process together with the disease-specific features.

Glycoprotein Iib / IIIa is an integrin complex found on surface of platelets. It is a receptor for fibronectin, fibrinogen, plasminogen, prothrombin, thrombospondin and vitronectin. Its activation leads to platelet/platelet interaction via binding of soluble fibrinogen consequently causing prompt platelet aggregation.

Aim of the Work

Estimate the plasma level of soluble GpIIb / IIIa in β- thalassemia major patients.

Subjects and Methods

The study was conducted on a total of 67 individuals divided into two groups. The first group included 47 thalassemia major (TM) patients, while the second group included 20 healthy volunteers as control subjects.

Patients with other forms of thalassemia were excluded, as well as Inflammation and infection. All subjects in the current study were subjected to thorough history taking with emphasis on transfusion history and chelation therapy, complete physical examination including spleen, liverand routine laboratory investigations; Complete blood count, serum ferritin, CRP, and measurement of plasma level of GpIIb / IIIa by ELISA. Calculation of thrombosis risk was done using thalassemia related thrombosis risk scoring system TRT-RSS.

Results

Table 1: Comparison between the three studied groups according to GpIIb / IIIa, Ferritin, and CRP.

	Cases		Control (n = 20)	Test of Sig.	p
	Non-splenectomised (n = 22)	Splenectomised (n = 25)			
Ferritin (ng/mL)					
Min. – Max.	600.0 – 6250.0	821.0 – 11192.0	30.0 – 80.0	H= 41.780*	<0.001*
Mean ± SD.	2701.2 ± 1684.2	3202.6 ± 2372.4	51.00 ± 14.35		
Median (IQR)	2223.5 (1674.9 – 3613.0)	2650.0 (1950.0 – 3229.0)	50.00 (39.0 – 59.50)		
Sig. bet. grps.	p_1=0.579, p_2<0.001*, p_3<0.001*				
Gp2b/3a (ng/mL)					
Min. – Max.	2.94 – 11.88	3.63 – 21.11	0.04 – 3.92	H= 46.721*	<0.001*
Mean ± SD.	5.24 ± 2.04	9.20 ± 4.33	2.30 ± 1.05		
Median (IQR)	4.98 (3.75 – 5.88)	8.05 (5.87 – 9.83)	2.16 (1.43 – 3.25)		
Sig. bet. grps.	p_1=0.003*, p_2<0.001*, p_3<0.001*				
CRP (mg/dl)					
Min. – Max.	1.54 – 4.30	1.32 – 4.02	1.00 – 2.90	F= 9.925*	<0.001*
Mean ± SD.	2.74 ± 0.79	2.83 ± 0.82	1.92 ± 0.53		
Median (IQR)	2.89 (1.99 – 3.20)	2.90 (2.08 – 3.40)	1.95 (1.55 – 2.30)		
Sig. bet. grps.	p_1=0.901, p_2=0.002*, p_3<0.001*				

Table 2: Comparison between the splenectomised and non splenectomised groups according to TRT- RSS grade and TRT score.

	Cases				Test of Sig.	p
	Non-splenectomised (n = 22)		Splenectomised (n = 25)			
	No.	%	No.	%		
TRT- RSS grade						
Low risk	22	100.0	1	4.0	χ^2= 50.420*	MCp <0.001*
Int. risk	0	0.0	22	88.0		
High risk	0	0.0	2	8.0		
TRT Score						
Min. – Max.	0.0 – 6.0		8.50 – 17.0		U= 0.000*	<0.001*
Mean ± SD.	3.66 ± 1.59		11.88 ± 1.71			
Median (IQR)	4.50 (2.0 – 4.50)		11.0 (11.0 – 13.50)			

Conclusion

Thalassemic patients are characterized by stronger platelet aggregation as reflected by increased GpIIb / IIIa. The thalassemia-related thrombosis risk scoring system (TRT-RSS) is a useful risk assessment tool intended to predict initial thrombotic events in patients lacking previous events. Moreover, higher GpIIb / IIIa was associated with high , and intermediate TRT-RSS risk score.