Von Willebrand Factor in different collagen vascular diseases in pediatric patients

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Abstract

Von Willebrand Factor (vWF) is one of the circulating blood proteins that is produced and released by vascular endothelial cells and is frequently used as an indicator of endothelial damage and endothelial cell dysfunction in vascular disorders. Collagen vascular diseases (CVD) are characterized by abnormal immune system activity with inflammation in tissues as a result of circulating autoantibodies. Classic CVD in pediatrics include systemic lupus erythematosus (SLE), scleroderma, dermatomyositis (DM) and Juvenile idiopathic arthritis (JIA), formerly was known as juvenile rheumatoid arthritis (JRA).

Objective: **The purpose of the present study** was to identify the plasma level of von Willebrand factor antigen (vWF: Ag) % in different childhood onset collagen vascular diseases. Also to evaluate plasma vWF: Ag levels as useful indicator of collagen vascular disease activity.

Subject: We studied ٦٠ plasma samples from pediatric patients with collagen vascular diseases (٢٥ SLE, ٢٥ JIA and ١٠ DM). They were recruited from pediatric rheumatology outpatient clinic. ٢٥ children with SLE (٢١ girls – ٤ boys) aged ٤ to ١٢ years were sub grouped to ١٠ cases with active SLE and ١٥ cases with inactive SLE. ٢٥ children with juvenile idiopathic arthritis (١٥ girls – ١٠ boys) aged ١ to ١٢ years were sub grouped to ١٠ cases with active JIA and ١٥ cases with inactive JIA. ١٠ patients with active dermatomyositis (٨ girls – ٢ boys) aged ٥ to ٩ years. The control group constituted of ٢٠ control healthy children matched for age and sex.

Sample collection and analysis: The blood samples were collected in sodium citrated tubes from patients and controls. Plasma vWF: Ag was determined by enzyme-linked immunosorbent assay (ELISA) kit for the quantitative determination of von Willebrand Factor Antigen (vWF: Ag) in citrated human plasma (Helena Laboratories, Texas, USA) according to manufacturer direction

Results: The mean (SD) plasma vWF: Ag values in patients with active, inactive SLE and controls was found to be ١٩٩ (±١٩) %, ١١٣ (±٢١) % and ٥٥ (±٢٠) % respectively. The mean plasma vWF: Ag values of the patients with active SLE was statistically significant higher than in inactive SLE (P<٠.٠١) and controls (P<٠.٠١). In the patients with active, inactive JIA and controls, mean (SD) plasma vWF: Ag values were found to be ١٧٠ (±١٧) %, ١٠٩ (±٢٥) % and ٥٥ (±٢٠) % respectively. There was a statistically significant difference in vWF: Ag values between patients with active and inactive JIA (P<٠.٠١) and between active JIA and controls (P<٠.٠١). The mean plasma vWF: Ag values in patients with DM were found to be ٢٥٥ (±١٢٢) %. There was a statistically significant difference in vWF: Ag values between patient with DM and control group (P<٠.٠١).

Conclusion:

- Von Willebrand Factor increased during collagen vascular disease activity when compared with inactive disease and control.
- Von Willebrand Factor can be considered as a marker for collagen vascular disease activity. However, it is not specific marker.

Key Words: Von Willebrand Factor - Collagen Vascular Disease - Systemic Lupus Erythematosus - Dermatomyositis - Juvenile Idiopathic Arthritis - juvenile rheumatoid arthritis.