SEVERE VON WILLEBRAND'S DISEASE WITH ABNORMAL PLATELET AGGREGATION. H. Dohi (1, 2), K. Kobayashi (1, 2), A. Kawakami (1), K. Hara (1), H. Fujimura (2), A. Furutani (2), Department of Medicine and Pediatric, Kure National Hospital, Kure, Japan(1); Research Institute for Nuclear Medicine and Biology, Hiroshima Univ. Hiroshima, Japan(2).

A 17-year-old boy with a life long history of easy bruising, epistaxis, subcutaneous hemostases and prolonged bleeding time from minor injuries, was initially diagnosed as having von Willebrand's disease, who presented epistaxis at 2 years of age. Since his initial diagnosis, he has been treated with cryoprecipitate on many occasions to correct his bleeding tendency. The laboratory findings in the patient and his family are summarized in the table. The patient’s mother, father and one brother have not complained any bleeding tendency.

Bleeding time VIII:C $\pm$ 1.64); mean VIII:Rcof $\pm$ 4·5 100

Hypothroidism and acquired von Willebrand's disease

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A diagnosis of type IA von Willebrand’s disease was made in three patients presenting with a mild bleeding tendency. Previously unrecognized hypothroidism was also confirmed in two patients. In the third, hypothroidism was diagnosed four years after initial presentation. In all three patients, thyroid therapy was associated with correction of the haemostatic defect and resolution of the bleeding tendency. The association of von Willebrand’s disease and hypothroidism prompted us to examine the relationship between thyrocrin (TSH), thyroid and components of the factor VIII complex in 12 patients with clinical and biochemical hypothyroidism. Factor IX was also studied. Mean VIII:C (measured by 2 stage assay) was 0.90 u/ml (range 0.55 - 1.14); mean VIII: Ag 0.83 u/ml (range 0.44 - 1.64); mean VIII:Rcof 0.73 (range 0.45 - 1.53); mean factor IX 0.72 (range 0.39 - 1.19). Multimeric analysis of vWF:Ag performed in samples from 8 patients was normal. VIII:Rcof levels were significantly lower than those of normal controls. A significant inverse correlation was obtained between TSH and factor IX and T4 and vWF:Ag. Although there is a definite inverse relationship between TSH and factor IX, this is not evident with respect to factor VIII and a different mechanism is probably responsible for the modest reduction of vWF:Ag and the occurrence of clinically-evident von Willebrand’s disease which we have demonstrated in a small proportion of hypothyroid patients.

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