

HAIRY CELL LEUKEMIA: REPORT OF A CASE WITH A PLASMA MEDIATED DEFECT IN PLATELET FUNCTION AND PLATELET MORPHOLOGY. J.C. Mattson and S. Sanal. Department of Pathology, Michigan State University, East Lansing, MI, and the V.A. Hospital, Saginaw, MI.

A 60-year-old male with hairy cell leukemia developed purpura. Studies performed prior to splenectomy and at a time when purpuric lesions were present demonstrated a severe platelet functional defect characterized by absent primary and secondary wave aggregation with ADP and Ristocetin, absent Collagen aggregation, and diminished Epinephrine aggregation. In addition PF3 was decreased and the bleeding time prolonged. EM demonstrated a mixed population of degranulated, enlarged platelets and normal platelets. Following splenectomy PF3 levels and the bleeding time returned to normal. Repeat aggregation studied performed post-splenectomy showed return of primary wave aggregation but persistent absence of secondary wave aggregation in response to ADP, and virtually absent response to Collagen. Ristocetin aggregation returned to normal, but Epinephrine failed to produce any response. EM performed post-splenectomy demonstrated normal platelets. Tests performed on the patient's pre-splenectomy plasma indicated the presence of an inhibitor which was capable of inhibiting primary aggregation, nucleotide release and secondary wave aggregation in normal platelets with all agents tested. This inhibitor had no effect on adhesion in a collagen adhesion assay. Full inhibitory effect was present after incubation of inhibitor plasma and normal platelets for 5 min. but with longer incubation times inhibition of aggregation was less pronounced suggesting either dissociation or destruction of the platelet bound inhibitor. Partial characterization of the inhibitor demonstrated that it was dialyzable, stable at 56 C for 30 min. and at 4 C for 72 hrs., and present in a 50% ammonium sulfate fraction of plasma. These studies suggest that the inhibitor is a globulin which appears to exert its inhibitory effect by nonspecific, easily dissociable binding to the platelet surface.

CIRCULATING PLATELET AGGREGATES IN THROMBOTIC PATHOLOGIES. J.J. Rodzynek, P.L. Schoenfeld, T. Martin, P. Léautaud, P. Wettendorff and A. Delcourt. Department of Internal Medicine, Ixelles Hospital, Brussels, Belgium.

The dosage of circulating platelet aggregates following Wu and Hoak (CPA) was performed in 52 healthy volunteers acting as normal controls (N), in 205 consecutive patients admitted in a general hospital for pathologies other than thrombotic or cardiac ischaemia (group I), in 59 consecutive admissions for deep venous thrombosis (group II), in 45 consecutive admissions for pulmonary thromboembolism (group III), in 35 consecutive admissions for acute coronary insufficiency (group IV) and in 73 consecutive admissions for acute myocardial infarction demonstrated by electrocardiogram and specific enzymes (group V). The percentage of positive tests (CPA 0.80) in those various groups appeared as follows:

N	Group I	Group II	Group III	Group IV	Group V
3.8%	36.5%	52.5%	55.5%	40%	61.6%

In conclusion: The positivity of CPA is not a specific indicator of a thrombotic disease neither of acute coronary insufficiency (36.5% false positive tests). CPA is not sensitive for the diagnosis of thrombotic conditions. In the setting of ischaemic heart disease, CPA positivity appears with higher frequency when myocardial necrosis is demonstrated. However the two groups don't differ significantly on a statistical basis (P non significant).

PLATELET REACTIVITY IN CORONARY HEART DISEASE. J.M. Riddle, T.G. Lee, J. Rival, S. Goldstein, and P.D. Stein. Departments of Internal Medicine and Surgery, Henry Ford Hospital, Detroit, Michigan, USA.

Platelet reactivity was evaluated in 96 patients with coronary heart disease and 72 healthy subjects. Transmission electron microscopy was used in a standardized in vitro method to evaluate these platelets. The degree of surface activation (cytoplasmic spreading by single platelets) and aggregate formation were both recorded. A hyperactive response was defined as >20% of the spread type platelet and/or an increased number of aggregates (>93 aggregates/100 single platelets). Hyperactive platelet populations were found in only 8% (6 of 72) of normal subjects. In contrast, 46% (6 of 13) of patients with stable angina, 45% (10 of 22) of patients with crescendo angina, 67% (8 of 12) of patients with acute subendocardial infarction and 83% (40 of 49) of patients with acute transmural infarction showed hyperactive platelets. The mean percentage of the spread type platelet for the various coronary heart disease groups was 25, 26, 25 and 51 respectively with corresponding mean values of 101, 84, 71 and 123 for platelet aggregates. The reactivity of platelets from patients with coronary heart disease differed significantly from the normal subjects ($P < .01$). Patients with acute transmural infarction showed a significantly greater platelet reactivity ($P < .005$) when compared to the other groups of patients with coronary heart disease. Patients with all forms of coronary heart disease exhibited greater platelet reactivity than normal subjects. These data support the concept that increased platelet activity may be an important factor in acute transmural myocardial infarction and perhaps in coronary heart disease in general.

DISTURBANCE OF PLATELET FUNCTION IN THE NEPHROTIC SYNDROME. E. Walter, D. Deppermann, K. Andrassy, E. Weber. Medical Department, University of Heidelberg, Heidelberg, GFR

Thromboembolic phenomena often (30 %) complicate the nephrotic syndrome. It was therefore investigated, whether disturbed platelet functions play a role in this disease.

28 normals, 34 patients with nephrotic syndrome and 18 of them with impaired kidney function were tested. In 20 patients the measurements were repeated after administration of aspirin plus dipyridamole.

Patients with nephrotic syndrome showed in comparison to normals the following changes: 1. increased platelet count ($p < 0.01$), 2. enhanced platelet adhesiveness (Wright-test: $p < 0.001$), 3. increased spontaneous aggregation (PAT I: $p < 0.001$; PAT III: $p < 0.01$), 4. enhanced PF 4-activity (heparin neutralisation: $p < 0.001$), 5. elevated β TG-levels on -ly in impaired kidney function. There was no difference in the reaction of platelets against ADP as well as collagen. The changes in platelet function correlated with the severity of the nephrotic syndrome (proteinuria, hypalbuminaemia, hyperlipoproteinaemia). After aspirin plus dipyridamole administration spontaneous platelet aggregation and adhesiveness were normalized.

There is a disturbance of platelet function in patients with nephrotic syndrome, which can be reversed with antiaggregating agents.