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GLANZMANN DISEASE (THROMBASTHENIA) — A CASE REPORT

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INTRODUCTION

THE bleeding time is one of the tests of platelet functions. Quantitative or qualitative platelet dysfunction results in prolonged bleeding time. If the latter is normal further tests are not indicated (Cazapek, 1978).

Platelet control haemostasis by forming a plug at the site of injury and by promoting clotting mechanisms. It acts by adhesions, aggregations and release reactions (Arkel, 1976). One classification of the dysfunctions is given Hardisty (1974) which divides it into congenital forms such as thrombasthenia and defects of platelet release and the acquired variety which includes uraemia, paraproteinaemias or disseminated intravascular coagulation. Thrombasthenia (Glanzmann Disease) is restricted to cases with deficient A.D.P. induced platelet aggregations and deficient clot retractions (Hardisty, 1974). About 100 cases have been reported (Wintrobe, 1974). The rarity of this condition merits reporting this case.

CASE REPORT

An 18 year old male, admitted in April, 1978, with recurrent episodes of spontaneous bleeding on the face, forearms, legs, gums and prolonged bleeding after a trivial injury. He did not give any history of epistaxis, malaena or haematuria. All the other members of his family were normal. He did not take any drugs or anticoagulant nor did he suffer from any renal disease.

Examination revealed echymosis around the left eyelid and petechiae on the forearms and left leg. Hess's test was positive. No lymphadenopathy or hepatosplenomegaly was found. Laboratory results were a normal platelet count, a prolonged bleeding and partial thromboplastin time, impared ADP induced aggregations and no clot retraction. Platelet aggregations to collagen and isoprenaline were normal. Other tests of coagulations, renal, liver, disseminated intravascular coagulation were normal. The patient was transfused with fresh blood. The echymosis and petechiae disappeared within a few days.

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DISCUSSION

The patient with recurrent episodes of bleeding, had a prolonged bleeding time, despite a normal platelet count. Investigations indicated lack of ADP induced platelet aggregations but normal to collagen. There was no clot retraction. Impaired aggregations lead to inadequate thrombus formation and deficient platelet factor 3 (PF-3) availability. This results in impaired orientation to fibrin strands leading to deficient clot retraction (Hardisty, 1974).

There was no family history in this case. This disorder is inherited as an autosomal recessive and consanguinity is common in affected relatives (Pitmann, 1964). Spontaneous bleeding, though not disabling in our patient, was severe. Posttraumatic and post-operative haemorrhages may be serious (Wintrobe, 1974).

Contact activation of thrombasthenic patient's blood is subnormal. Tests which depend on contact activation such as partial thromboplastin time will be prolonged as in our patient. As this test measures the intrinsic pathway of blood coagulation, other tests which measure part of this pathway, for example thromboplastin generation test, may follow this test. This may cause a diagnostic error of Factor IX, Factor VIII deficiency (Wintrobe, 1974).

Platelet factor 3 availability was abnormal in the patient. Variable results occur (Cazpek 1978). Deficiency of fibrinogen is not a constant finding (Caen, 1966) and our patient has normal

level of fibrinogen. This condition is distinguished from other disorders of platelet dysfunctions which show a normal clot retraction and a collagen induced aggregation.

Drugs such as aspirin, which a common ingredient in many proprietary preparations, can inhibit platelet aggregations and release reactions. This may not be significant in a normal person. However, in patients with platelet dysfunctions or coexisting coagulopathy it may precipitate haemorrhage (Arkel, 1976).

SUMMARY

An 18 year old male admitted with recurrent episodes of echymosis, petechiae and bleeding gums. Investigations revealed a prolonged bleeding time, a normal platelet count, impaired ADP induced platelet aggregations and poor clot retraction. He was diagnosed as Glanzmann Disease and responded to fresh blood transfusions. This disorder may be mistaken for a disorder of coagulopathy. Surgery is hazardous. Drugs containing aspirin may precipitate haemorrhage.

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