Hemophilia A, Idiopathic Thrombocytopenia and HTLV-III-Infection Impressive Remission after Splenectomy:

A Case Report

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The simultaneous occurrence of hemophilia A and idiopathic thrombocytopenia with impressive remission after splenectomy is reported sporadically in the literature [1, 2]. In this report we describe a patient, who acquired HTLV-III-infection without signs of AIDS after treatment with factor VIII. Hepatitis B infection and other viral infections had been documented, too. In 1984 immune-thrombocytopenia was diagnosed so that HTLV-III and/or other virus infections may have caused thrombocytopenia.

Case Report

A 27-year old patient suffered, from birth, from a factor VIII deficiency, requiring substitution three times weekly. In addition, a brother and a nephew in his family suffered from hemophilia A. The patient is known to have chronic hepatitis B (anti HBc positive, anti HBs positive, and IgM HBc negative) since 1966. Hemorrhagic complications occurred in 1972 with a subdural hematoma, and in 1977 synovectomy in the region of the right ankle joint after a joint bleeding occurred.

In September 1984 petechiae appeared for the first time, concurrent with thrombocytopenia of 18,000/ μ l. The bone marrow showed an increase in megakaryocytes. The simultaneous proof of an IgG load of the autologous thrombocytes allowed a diagnosis of autoimmune thrombocytopenia. No free serum platelet auto- or allo-antibodies were found in our patient. After immuno-suppressive treatment with Decortin (starting with 100 mg/day) an increase in thrombocytes 178,000/ μ l was obtained. In March 1985 a renewed thrombocytopenia showed no remission after treatment with high-dose Decortin and additional Imurek. In July 1985 the patient was hospitalized with herpes zoster on the right leg and a painful lymphadenopathia in the right inguinal region. We found a positive proof of herpes zoster (titer 1:512), herpes simplex (titer 1:32), and cytomegalia (titer 1:32). The thrombocytes were reduced to 7,000 μ l. Repeated treatment with Decortin and Imurek showed no therapeutic success, and even the application of high-dose gamma globuline (Sandoglobulin 30 g/day for 5 days) revealed no increase in thrombocytes. In September 1985 a splenectomy was performed without complications, with

The pathogenesis of chronic immuno-thrombocytopenia is not yet clear. In our patient, the chronic form of an idiopathic thrombocytopenia could be substantiated by the proof of platelet-bound IgG, lack of significant splenomegalia, and shortening of survival time of thrombocytes. In the treatment of immuno-thrombocytopenia, glucocorticoids are first-line therapy. In therapy-refractory cases either the application of immuno-suppressiva or infusions with immuno-globulin are chosen as promising results have recently shown [5,7]. In our patient glucocorticoids and immunosuppressiva only led to a transitory increase in thrombocytes; a treatment with immunoglobulin over 5 days also showed no therapeutic effect. Only when a splenectomy was

performed after 12 months of unsuccessful immunosuppressive treatment, did this result in an impressive increase in thrombocytes. The patient is now still in remission.

An IgG-load of autologous thrombocytes, with concurrent thrombocytopenia, was described for different diseases (e.g. liver affections and viral infection) by Mueller-Eckhardt et al. [3]. Our patient exhibited positive antibody titers against herpes zoster, herpes simplex and cytomegalia. He had also been suffering from a chronic B-hepatitis since 1966. Thus, the possibility exists that the chronic hepatitis B as well as previously endured virus infections may have caused the development of the idiopathic thrombocytopenia. On the other hand, our patient showed a chronic HTLV-III-infection, confirmed by ELISA and immunoblot technique, without signs of immu-

Table I. Lymphocyte subpopulations in the peripheral blood of a patient with hemophilia A and idiopathic thrombocytopenia

3/85
4/85
5/85
Normal range (cells/ μ l)
Leu2
453
1,341
1,459
350- 840
Leu3
772
1,014
681
650-1,180
Leu4
2,425
2,978
2,604
1,240-2,120
L3/L2 Index
0.54
0.78
0.47
1- 3

nodeficiency (table I), a constellation frequently found in patients with hemophilia [4]. It has been discussed by Stricker et al. [12] that this retrovirus may play a role in the development of autoimmune ITP. It is known from the literature [13] that virus infections may also result in immuno-thrombocytopenia. In our case, therefore, it remains unclear, which kind of virus may have induced the thrombocytopenia.

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