Glanzmann Thrombasthenia Successfully Operated for Nasal Deformation with Recombinant Factor VIIA

An 8-year-old boy diagnosed as Glanzmann thrombasthenia (GT) by other center was referred to our department in March 2005. He has been followed with GT for 4 years. He complained difficulty in breathing and recurrent upper respiratory tract infections. We observed a big perforation at nasal septum and granulation tissue related with forgotten nasal tampon on nasal examination. Results of complete blood count were as follows: hemoglobin 8.9 g/dL, mean corpuscular volume (MCV): 64 fl, white blood cell count 4800/mm^3, and platelet count 321000/mm^3. He was hospitalized for cleaning of the granulation tissue. In our patient alloimmunisation had developed due to frequently platelet transfusion to stop resistant nasal and gingival bleedings before the admission. The patient was operated with local anesthesia. Forgotten tampon was removed and granulation tissue was resected. Regeneration of septum nasi was performed. Bolus injection of Recombinant activated factor VIIa (rFVIIa) 90 μg/kg was given immediately before operation and three times with 2 hours intervals after the surgery. The patient was discharged one day after without any complication.

Clinical efficacy of rFVIIa in thrombasthenic patients is not clear. Thrombin generation is impaired in GT patients. The ability of high-dose rFVIIa to improve thrombin generation through direct binding to activated platelets and/or overcoming the inhibitory effect of zymogen FVII may contribute to its therapeutic efficacy in GT patients(1). rFVIIa is postulated to act on platelets to activate factors IX and X and thus enhance thrombin generation. Other experimental work has suggested that rFVIIa can restore platelet adhesion defect by tissue factor-independent rFVIIa-mediated thrombin formation(2). rFVIIa given as bolus injections appears to be a safe and mostly effective alternative to platelet transfusion for the treatment and prevention of bleeding in patients with GT, particularly for those with antiplatelet antibodies and/or refractoriness to platelet transfusions.

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