Surgical treatment of multi-localized pseudotumor in patient with hemophilia A and an inhibitor: clinical case.

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OBJECTIVES

•One of the most often manifestations of hemophilia are muscle hematoma, less often we meet with subperiosteum and intraosseous localizations. When the treatment is deferred or not full such hematoma can be transformed into pseudotumors. The most severe complication of pseudotumor is its infection and spontaneous dissection with destruction of underlying tissues. That can be accompanied by massive uncontrolled bleeding especially in hemophiliac patients with an inhibitor. Surgical extirpation of pseudotumors usually is technically difficult and can be complicated with bleeding. Nowadays we meet pseudotumors rarely due to current opportunities of hemostatic therapy.

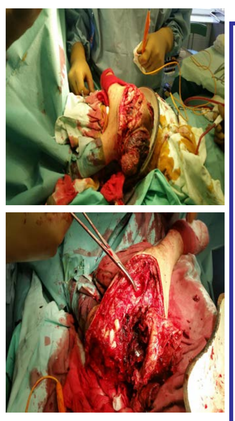
METHODS

•Anamnesis morbi: first sings of pseudotumor about 5 years ago, when first tumor formations appeared in left elbow and right foot and began to grow slowly. Factor VIII therapy was occasional without significant effect. In January 2015 his health changed for the worse when in the hospital in his country elbow pseudotumor was punctured. After that the pseudotumor increased in size. In august 2015 pseudotumor spontaneous dissection and long unstoppable bleeding occurred. Patient A., age 17, with hemophilia was hospitalized in Hematology research center, Moscow, Russia with fever up to 40° Celsius. The patient had a giant pseudotumor on right arm from middle shoulder to middle forearm with multiple skin necrosis, sings of proceeding bleeding and pseudotumor infection. More to it the patient had one more pseudotumor located on the right foot, sings of severe anemia (Hb=67 g/l). The patient was from one of neighboring to Russia countries, where he was offered an amputation as the only treatment.

•Bacteriological examination of right arm pseudotumor discharge: Escherichia coli, Klebsiella pneumoniae, Enterobacter cloacae, Staphylococcus aureus, Enterococcus faecalis.

•The patient had an inhibitor up to 50 BE in blood.

Due to MCT of the arm: proximal part of ulnar bone and olecranon were replaced with pseudotumor. Bone structure of radius and humerus was not changed. MCT of the foot: anterior part of talus is replaced by pseudotumor, cuboid totally replaced by pseudotumor, head and body of V metatarsal partially destroyed.



RESULTS

•Surgical treatment – extirpation of the pseudotumors of arm and foot - was performed on hemostatic therapy Eptacog a activated (Koagil VIIa) 120 mkg/kg every 2 hours. The surgery lasted 4 hours, blood loss – 2000 ml (patient`s weight 40 kg). After surgery there was massive infusion-transfusion therapy. In postoperative period hemostatic therapy Koagil VIIa, antibacterial therapy due to bacterial sensitivity and anemia correction continued. The wounds were left opened with antiseptic napkins and vacuum drainage system. The function and movement ability were saved both in arm and foot.

CONCLUSIONS

•Contemporary progress in surgery and hemostatic therapy allows to perform successful treatment of massive pseudotumors with bone destruction and infection. Surgical treatment of hemophiliac pseudotumors should be carried out in special medical centers, where hemophilia is a known disease.

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