MULTIPLE SEROUS, MUSCULAR AND SUBCUTANEOUS HEMORRHAGES IN A PATIENT WITH HEMOPHILIA A (a case report)

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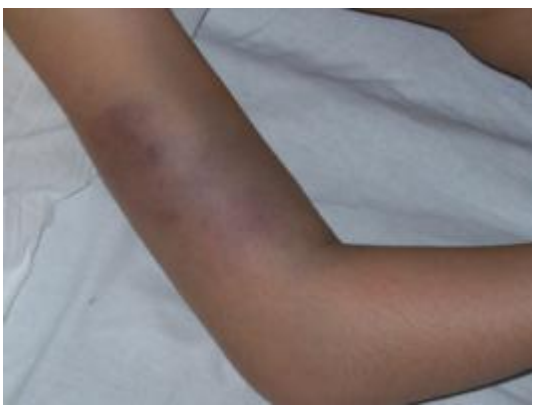
Hemophilia A is a X-linked bleeding disorder caused by absence or deficiency of coagulation factor VIII. However, 30% of the cases occur as a spontaneous mutation, and therefore, there is no family history of the disease. Hemophilia A is classified according to the baseline plasma level of FVIII into severe (factor level lower than 1 U/dL or less than 1%), moderate (factor level between 1 U/dL and 5 U/dL, or between 1% and 5%) and mild (factor level higher than 5 U/dL or greater than 5%). This classification system serves as a guide to the expected frequency of bleeding (1). The most common clinical manifestations of the disease are hemarthrosis and muscle-joint hematomas. In the literature are reported single cases of massive hemorrhages in serous cavities. Pulmonary and pleural bleeding are uncommon complications in hemophilia. Massive hemothorax and hemoptysis are rare. Intra-abdominal or retroperitoneal bleeding is far more common in hemophiliacs than are the usual abdominal emergencies (2).

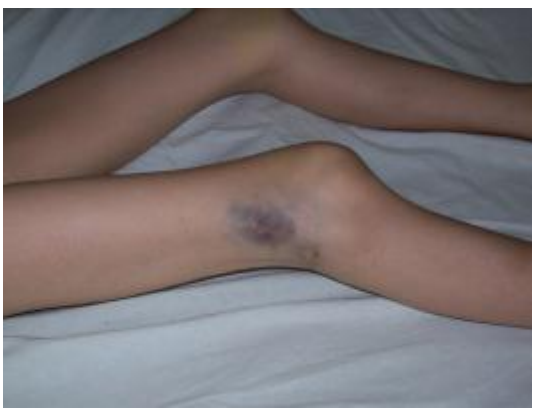
We present a child with moderate type of hemophilia A with clinical and image data for hemothorax, hemopericard, hemoperitoneum, multiple muscle hematomas and hemarthrosis of left elbow and right hip joint, as a result of minor trauma in the abdomen.

T. S. I. 10 years old boy, with moderate hemophilia A (Factor VIII 3%) from infancy. With multiple hospitalizations in Pediatric department – Dobrich and Pediatric clinic of Oncohematology – Varna, for relapsing hemarthrosis, single retroperitoneal hematoma and gastrointestinal hemorrhage. He did not perform prophylaxis with factor VIII.

The child was admitted in the Pediatric clinic of Oncohematology in September 2005 because of fever, abdominal pain and nausea for 1 day. Two days ago started pain in the right elbow and right hip. The complaints are connected with trauma in the abdominal region and right hip joint.

General condition: severe condition, subfebrile 37,5 gr.C. In compulsory sitting position. Pale skin and mucoses. Gracile habitus. Micropolyadenia. Head and neck – normal. PS – vesicular breathing, lowered in the bases. HVS – rhythmic heart rate, clear tones, HR 120/min. Abdomen – painful under palpation, marked in the left abdomen, which is edemic and warmer. Subcutaneous hematoma in the umbilical and periumbilical region. Liver and spleen – not enlarged. Extremities – limited movements in right elbow with intra and periarticular hematoma; hematoma on the right hip in the upper – lateral part of the hip with extremely limited movements of the right hip joint. Hypotrophy of the muscles of the right leg. Succusio renalis – negative. Mouth – spontaneous gingival bleeding. Fig. 1, Fig. 2



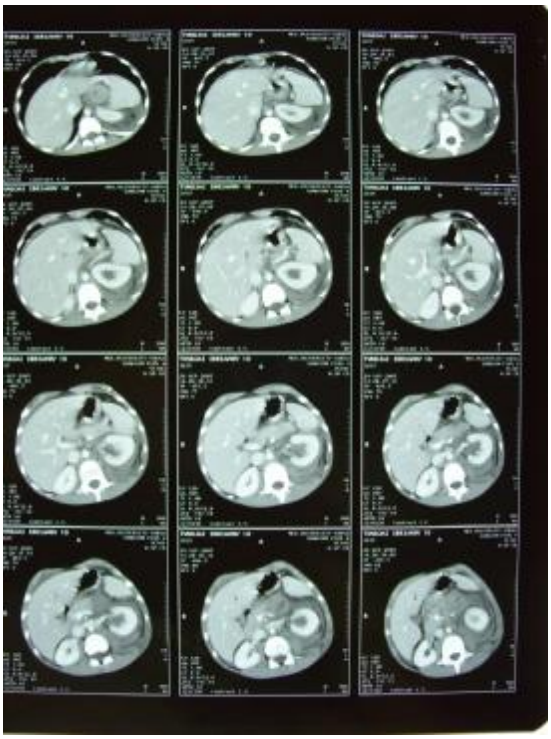


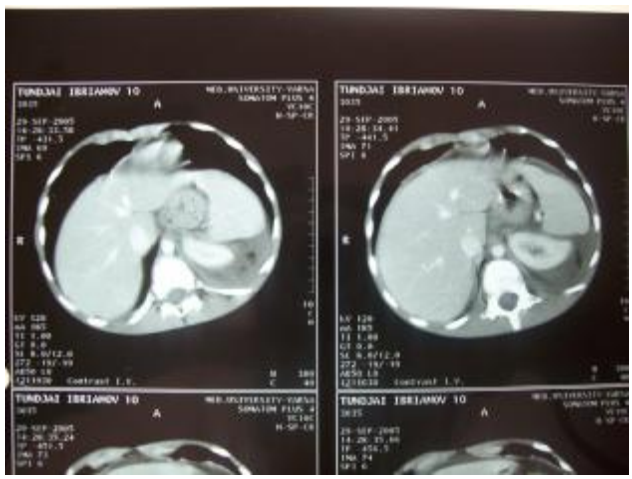
Laboratory findings: Hb 66 g/l, leuc. 23,4.10^9/l, Tr. 381.10^9/l. Bleeding time 120", clotting time over 600". Biochemical indexes – normal. CRP 33,6 mg/l, Protrombine activity 69 %, INR 1,6, APTT 79". Urine – albumin ++, sediment – 5-6 leuc., crystals uric acid, α amylase 137 U/l.

Ultrasound sonography: Spleen **–** homogenous, normal echogenity with normal capsule. Left kidney – hydronephrosis I grade, normal structure. Free liquid in the abdominal cavity (intraperitoneal).

Consultation with a surgeon: Abdomen – symmetric, on the costal level, spare no breathing, diffuse palpatory pain, marked in the left abdomen; local defance in the left subcostal region. Physiological peristaltic, stupid percussion tone: data for free fluid in the peritoneal cavity (probably blood), proved with ultrasound sonography. In conclusion: Hemoperitoneum, suspicion for two-moment rupture of the spleen and hemoretroperitoneum.

Abdominal CT №3035/29.09.05: Dilatated and firm tissues of the left abdomen. Liver – not enlarged, homogenous, normal vessels. Spleen – not enlarged, homogenous. Presence of free fluid with high density /30-40 HU/ in the abdominal cavity and pelvis. The biggest amount of blood is found in the left hypohondrium around the spleen and left kidney. LK – enlarged, with thickened parenchyma and hydronephrosis in the pelvis. Thickened left fascia Gerota. Firm and blooded pararenal fat tissue and thickened left musculus psoas. The changes in the dorsal region invade part of the paravertebral muscles. Free fluid with characteristic of blood is visualized on the length of left paracolic and infracolic space and in front of the big abdominal vessels, reaching the pelvis, where stratifies the intestinal stitches and is settled mainly in the left abdomen. Dislocated urinary bladder. Fig.3, Fig. 4.

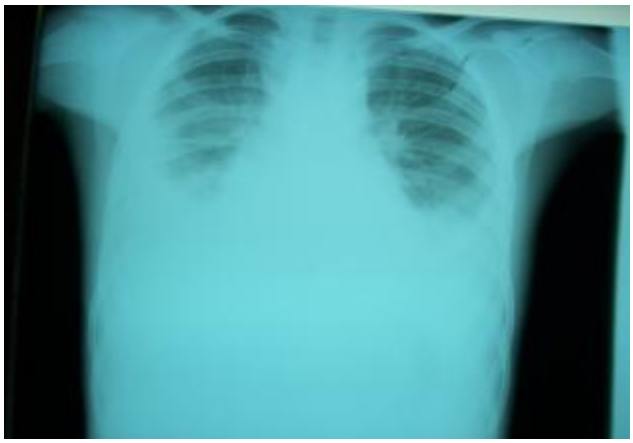




The treatment was started with Factor VIII in dose 2 x 600 U /50 U/kg./, Zinacef, Gentamicin, Metronidasol, blood transfusion, painkillers.

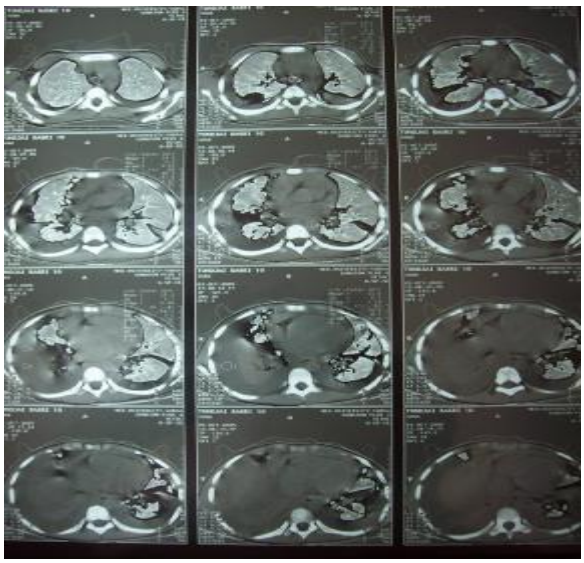
On the second day, after the hospitalization, was registered worsening of the general condition including epistaxis, hardened breathing, chest pain, increasing paleness, protruding jugular veins, tachipnea 48/min., tachicardia 120/min., BP 100/60 mm and auscultation data for lowered vesicular breathing, mainly in left subscapulary. The abdominal circumference increased (from 61

sm in admittion to 64 sm), adequate diuresis. From the laboratory tests - Hb 55 g/l, APTT 99". Roentgenological data for bilateral pleural exudation Fig. 5:

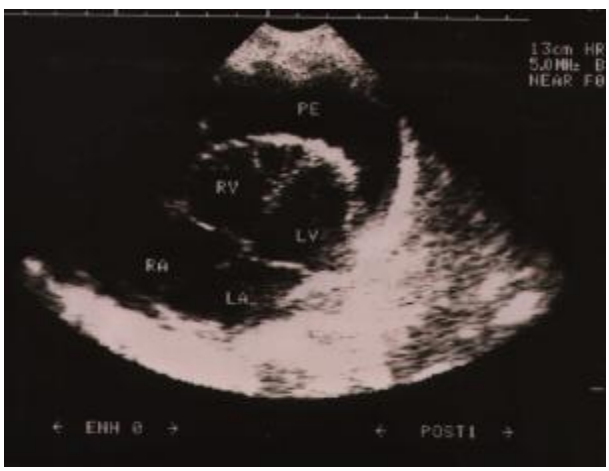


On the control **abdominal ultrasound and CT** there was no evidence of increased intra/extraperitoneal fluid.

In the native chest CT scan were found: bilateral pleural exudates with roentgenologic density – 30-40 HU (Hemothorax) with compression of the right pulmonary parenchyma from the exudates. There is a small collection in the pericardium with high density over 30 HU – Hemopericard. Fig. 6:



EchoCG – structurally normal heart. Normal venous drainage. Intact IAB and IVB. RV – 14mm, LV 39/23 mm, FS 41%. Evidence of **pericardial exudates**. Fig. 7:



The treatment was continued with single application of Factor VIII in dose 2400 U (100U/kg), followed by 24 hour intravenous infusion of 2400 U and Solumedrol 2 x 20 mg. The total duration of the replacement treatment was 16 days, as in the last six the doses were reduced to 2 x 600 U.

During the treatment the general condition of the child gradually improved with seizure of the clinical symptoms of respiratory insufficiency and subjective feeling of chest pain. The objective examination of the abdomen showed stable dynamics of reduction of the free fluid (blood) and lowering of the abdominal circumference to 54 sm. The image tests (roentgenological follow-up and abdominal ultrasound) of the hemorrhages in the bodily cavities, showed stable dynamics, for lowering and full disappearing, till the end of the hospitalization. Fig. 8, Fig.9

Fig. 8.

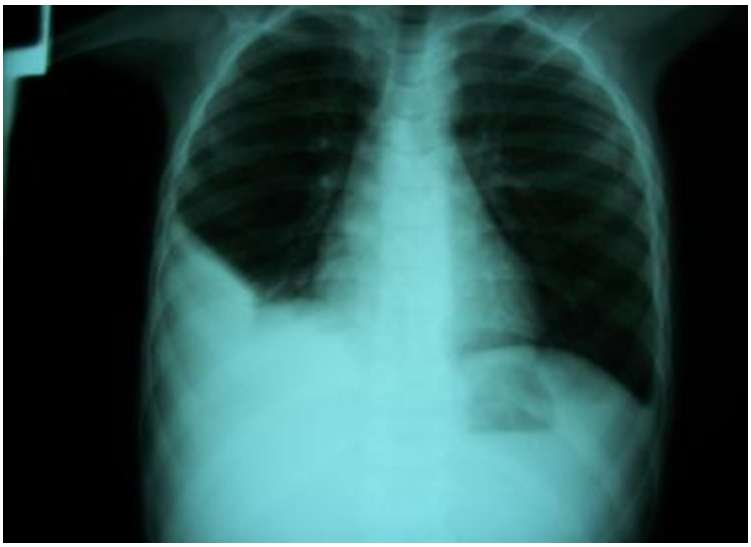
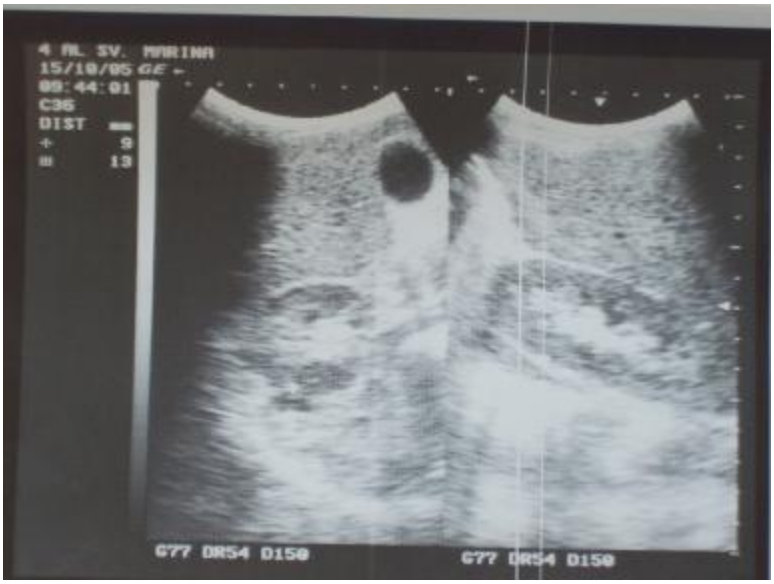


Fig. 9.



After the dehospitalization from the clinic, till now, the child is on a prophylactic regimen with factor VIII once a week in a dose 20 U/kg. During the three years follow up, the patient was hospitalized six times because of relapsing posttraumatic hemarthroses on both knee joints.

In conclusion, the reported case is a casuistic with multiple hemorrhages, in a child with moderate type of Hemophilia A, mastered with continuous infusions of high dose Factor VIII. This clinical case is an approval to the fact that in the criteria for starting prophylactic regime, despite the level of Factor VIII, the severity of the clinical manifestation must be taken in mind.

References:

1. Price VE, Hawes SA, Chan AKC. A practical approach to hemophilia care in children. *Paediatr Child Health* 2007;12(5):381-383
2. Charles S. Davidson, Robert D et al. Hemophilia. A clinical study of forty patients. *Blood* 2007;1949 (4):97-119