ONE CASE OF NEUROLOGICAL COMPLICATION WITH GIGANTIC CAVERNOSAL HEMANGIOMA — SYNDROME OF KAZABACH-MERRY

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The syndrome of Kazabach-Merry is a disease which is included in the Classification of haemorrhagic diateses, the group of coagulopathies, i. e. innate (congenital) vasopathies. It was reported first by Kazabach and Merry in 1940: congenital gigantic cavernosal hemangioma on the lips' skin, shoulders, head, abdomen, buttocks, thighs, etc. which with the time grows bigger and begins bleeding. The hemangioma is detected in the early childish age. In the past the disease was named «hemangioma-thrombopenia». Heglin described «consumptive coagulopathy» which shew disseminated intravasal coagulation and defibrinating syndrome. With the age the disease becomes heavier. Possible stomach, intestines, brain and other organs' haemorrhages with additional heavy anaemia are reported.

We report one our case in the Clinic of Neurology and Neurosurgery which was pathoanatomically verified: H. H. H., age 58, from Chernook village, District of Varna; clin. record № 22 193/1979. On 22nd September, 1979 (12,00 a. m.) the patient had an immediate weakness of lower extremities and thinking that it was a fatigue he went to bed. Two hours later he woke totally immobile with both lower extremities. He reported that he had since birth «tumour-like» formations scattered on the skin, lips, back, thorax, abdomen and right inguinal place; the formations are similar to varicosal knots. Somatic examination: similar to lipomas formations are established on the skin; at certain places they represent conglomerates and convolutes of vessels with honeycomb structure. Cor et pulmo: normal status, RR 125/80. Liver is palpated 4—5 cm under costal arch, oval edge, no palpatory pain. Spleen — normal.

Neurological status: lower slight paraplegia, knee and Achiles reflexes are not detected bilaterally, also abdominal reflexes bilaterally. Total anaesthesia for profound and superficial sensitiveness from conductive type at L-1 and L-2 dermatoms downward. Expressed muscle hypotonia of both legs. The patient has retentio urinae et alvi.

Paraclinical (laboratory) data: Hb — 9,5 g %; later it is 7,6 g % even 5,8 g %; erythrocytes — 2 330 000 and 1 900 000; leucocytes — 8800 and 11 800; DBC — normal distribution; sedimentation test (Panchenko) — 12 mm, Westegreen — 10/15 mm; hematocrit — 20,26; blood urea — 42 mg %; blood sugar — 103 mg %. Liver tests: Weltmann — 5, 4 and 6 TT; thymol — 0,28 meKv; ionogram — normal; direct bilirubin — 1,26 and indirect — 0,63; alkaline phosphatase — 49. Urine: protein-positive; sediment — 15—20 leucocytes. Coagulation status: time of bleeding — $1^1/_2$ min, time of coagulation (clot-formation) — 8 min; thrombocytes — under 200 000 in mm^3 with last values 100 000—120 000; megathrombocytes are detected; fibrinogen —

between 28 and 140 mg % (normal value 200—400 mg %). Prothrombin time — between 40 and 70%; cephalin-caoline time — 52 and 63 sec.; recalcification — 97 and 180 sec; thrombelastogram: ER — $5^1/_2$ min; Ka-coagulation time — 15 min and 23 sec (elongated); MA — maximum amplitude — 15 mm.

Liquor diagnostic: first lumbal puncture was applied at the level of L-4 and L-5 (29th Sept., 1979) — strongly xanthochromic liquor was collected. Masses of old and fresh erythrocytes with protein content 1680 mg % were registered. Liquor was curdled as if an egg white. Second puncture (20 days later) — liquor was slightly xanthochromic with protein 80 mg %. Last liquor (10th November, 1979) was normal with protein 20 mg % and transperant colour.

X-rays diagnostics: the contrast material was stopped for a while at Th-10 and Th-11 with suboccipital myelography. Conglomerate of pathologic vessels can be detected on cyliacography between 10th and 11th rib (from the right); the vessels are filled via the directin of the left branch of arteria hepatica. Venecavography shows a thrombocytic lower vena cava. With all native X-rays photos can be seen dense round phlebolites participating in the gigantic hemangioma. After a consultation with gastroenterologist we suggested a hepatic cirrhosis (minding that $^1/_3$ of the cirrhosis preserves its normal biochemical indexes — similar to our case). Biopsy was applied for a histological study. The skin was opened and multiple connected or single honeycomb-like cavities (similar to echynococcal cysts) filled with fibrous prominences were found under it. Clots and thrombs, also haemolysated blood was established after the cavities were cut. Some of the thrombs were hialinysated. The neurological complication in our case was performed as a spinal insultus (haematorrhagies, haematomyelia) begun after haemorrhages of the vessel malformations located intrarhachially and extradurally.

The treatment included haemotransfusion (blood-group A1B), fibrinogen, urbason, ceporex, venoruton, nelidix, vitamin C. Regardless of the applied medicines the patient developed chroniosepsis due to the heavy decubituses, constant catheter, urea content-increase, hypertonia and fibrility. The heavy coagulopathy and anaemia were the reason for a progressive change for the worse; with cardial and pulmonal insufficiency on 11th December, 1979 the patient died.

According to Dimitrov (1) the Syndrome of Kazabach-Merry begins its development as a congenital vasopathy and with time it is formed as a combined haemorrhagic diathese — angiocoagulothrombopathy. R. Heglin (5) accepts that every intravasal clot affects thrombocytes, fibrinogen and rest coagulating factors which after that are consumed for the coagulation itself. He reports fibrinous monomers in plasma. According to Stubert (after Dimitrov) there are some cases with decreased production of thrombocytes in the bone-marrow due to a process of retension in megacariocytic apparatus. Goot (after Majdrakov) suggests that the gigantic innate haemangiomas with massive destruction of blood-plates induce the production of autoimmune antibodies directed towards megacariocytes and thrombocytes themselves.

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ОБ ОДНОМ СЛУЧАЕ НЕВРОЛОГИЧЕСКОГО ОСЛОЖНЕНИЯ ГИГАНТСКОЙ КАВЕРНОЗНОЙ ГЕМАНГИОМЫ — СИНДРОМ КАЗАБАХ—МЕРИ

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РЕЗЮМЕ

Авторы описывают случай неврологического осложнения гигантской кавернозной гемангиомы — синдром Казабах—Мери. Гематорахия (соответственно гематомиелия), которые наступили при кровоизлиянии малформаций сосудов, расположенных интрарахиально и экстрадурально, привели к спинальному инсульту.