Clinical case: Syndrome of cutis marmorata teleangiectatica congenital with multiple ulcers in a newborn child

**Keywords:** cutis marmorata teleangiectatica, marbled skin, teratogenic factors, human beta-chorionic gonadotropin, Klippel-Trenone syndromes, genitourinary system malformations, hydrocephalus, glaucoma

**Clinical report**

Syndrome of cutis marmorata teleangiectatica congenital (CMTC), also known as Cato van Lochuizein syndrome, is a persistent patchy skin lesion. The syndrome was first described in 1922 by Dr. Cato van Lochuizein. More known benign transient syndrome of newborn “marbled skin” differs from the CMTC by less firmness and intensity of the skin process. Subsequently, more than 300 cases of the disease have been described in the world literature. The etiology and pathogenesis of the CMTC still unknown. It is assumed that there is a association with teratogenic factors, an increase of the human beta-chorionic gonadotropin level, autosomal dominant inheritance with incomplete penetrance is not excluded. A combination of the CMTC with Sturge-Weber and Klippel-Trenone syndromes was also noted. A number of authors had included all three syndromes in a group of vascular diseases associated with mesodermal system malformations. In support of this theory, concomitant asymmetry of the body (hyper or hypotrophy of the extremities, syndactyly) and the fact that in 50-60% of cases CMTC is associated with cardiac anomalies, genitourinary system malformations, hydrocephalus, glaucoma with other accompanying vascular malformations.

It is known that CMTC can be presented by vascular spots, telangiectasias of various intensity of red color, up to areas of skin atrophy, followed by ulceration and scar formation. The intensity of the spots depends on the air temperature - in the warm they become lighter.

Histopathology of the syndrome: According to Kienanst and Hogeher, the diagnosis of BMTC is based on the main and secondary clinical criteria.

The main criteria are: congenital marble erythema, which does not disappear during rubbing and heating and does not have venous ectasia.

Secondary criteria:

I. Telangiectasia
II. Ulceration
III. Atrophy within the focus
IV. Easy palliation during the first years
V. Nevus flammeus or Mongolian spot (on another site)

CMTC is more often localized at limbs, lower more often, rare –trunk and face. Despite the variability of the course of CMTC, in most cases, skin lesions undergo regression to adolescence.

The cases of combination of CMTC with developmental anomalies, as a rule, require syndrome treatment.

Aim: Description of the case of observing the child with a syndrome of cutis marmorata teleangiectatica congenital.

Born on 23.03.17, boy T. was referred on 24.03.17 from the regional hospital to the children’s republican clinical hospital (Kazan) in a severe condition with the diagnosis: “Intrauterine infection. Intrauterine pneumonia? Necrosis of subcutaneous fat?” Severity of the condition due to to respiratory failure, neurologic symptoms, skin syndrome.

Child was born full-term on a gestational age of 40-41 weeks, gravida 5 para 5, the pregnancy without pathology. The mother was under observation due to HIV infection (R75 code for ICD). Body weight at birth was 3280g, height 51cm, Apgar score 7-8, with asphyxia episode. In the maternity ward, infusion therapy and antiretroviral therapy (retrovir medication, a dose of 4mg/kg) were performed.

A survey plan was drawn up: a hematology test, a biochemical blood test, a blood test for the acid-base state, a coagulogram; dopplerography of the inferior vena cava, iliac veins, echocardiography, neurosonography, ultrasound examination of the abdominal cavity, dermatologist consultation. Child showed restless behavior, moderate muscular hypotension, decreased unconditioned reflexes, subichetery of the skin, moderate cyanosis of the nasolabial triangle.

Local status: In the right iliac region and on the right lower limb (thigh, drumstick, foot), there were moderate edema of the soft tissues, segmental lipodystrophy with 0.8 to 1.5cm foci, as well as multiple different sizes and configurations purple-violet dark spots (the “marble” vascular pattern), with clear boundaries; painless; not disappearing with diascopy and warming. In the region of the knee and shin, three foci of ulceration with diameters of 0.4 m, 0.8cm and 1.2cm, (Figure 1). The skin outside the lesions was warm, with a pale pink coloration. Central vessels pulsation is normal. Soft tissues hypotrophy of the right lower limb with normal length was noted.
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**Acknowlegements**
None.

**Conflict of interest**
Authors declare there is no conflict of interest.

**References**


