

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.¹ 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.^{2,3} 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.^{4,5} A combination of the CMTC with Sturge-Weber and Klippel-Trenone syndromes was also noted.⁶ A number of authors had included all three syndromes in a group of vascular diseases associated with mesodermal system malformations.⁷ 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.⁸⁻¹²

It is known that CMTC can be represented by vascular spots, telangiectasias of various intensity of red color, up to areas of skin atrophy, followed by ulceration and scar formation.¹³ The intensity of the spots depends on the air temperature - in the warmth they become lighter.¹⁴

Histopathology of the syndrome: According to Kienanst and Hoeger,³ the diagnosis of BMTC is based on the main and secondary clinical criteria.³

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.¹⁵ Despite the variability of the course of CMTC, in most cases, skin lesions undergo regression to adolescence.³

Volume 4 Issue 3 - 2018

Nurmeev ¹ N,¹ Nurmeeva A R²

¹Kazan Medical University, Russia

²Republican Clinical Dermato-Venerology Dispensary of the Ministry of Health of Republic of Tatarstan, Russia

Correspondence: Ildar Nurmeev, M.D., professor of pediatric surgery department, Kazan State Medical University, Butlerova Street, 49, Kazan, Russian Federation, 420012, Russia, Tel 00 7843 2678 969, Email nurmeev@gmail.com

Received: April 19, 2018 | **Published:** May 17, 2018

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 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, subicheterity 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 hypertrophy of the right lower limb with normal length was noted.



Figure 1 Child T., 10 days old. Cutis marmorata telangiectatica congenital, right lower limb photograph

- A. Brain ultrasound: Tissue hypertension syndrome and hypoxic-ischemic injury signs.
- B. Hematology: neutrophilic leukocytosis with a left shifted leukocyte formula.
- C. Neurology: hypoxic-ischemic injury of the central nervous system. No association with TMPC have been identified.
- D. Ultrasound of the vessels of the limb and examination of the vascular surgeon: pathological changes not revealed.
- E. Heart ultrasound: without clinical significant disorders.
- F. Ultrasound of the abdominal cavity organs: pathology was not revealed.
- G. Intrauterine infection was not confirmed.
- H. Differential diagnosis had been carried out with marbled skin in newborns. Last one condition is referred to as physiological, disappears spontaneously and only occasionally has a persistent character. In case with ulceration, diagnosis also requires exclusion of purulent-inflammatory diseases of soft tissues.
- I. Follow-up monitoring of the child demonstrated a favorable course of the disease. All areas of necrosis were closed by healing with skin scarring. The sites of lipodystrophy were transformed into foci of atrophy of subcutaneous fat.

Conclusion

The case of cutis marmorata telangiectatica congenital in a newborn with localization on the right lower limb and right iliac region was detected immediately postpartum and was accompanied by ulceration, soft tissues atrophy of the right lower limb. Examination did not reveal abnormalities of the heart, abdominal organs and vascular system.

Clinical severity with ulcerative necrotic lesions of the skin and subcutaneous fat and limb hypotrophy was the point of difficulty of differential diagnostic search. The clinical case is described due to rare occurrence of the disease with a severe clinical course of the skin syndrome.

Acknowledgements

None.

Conflict of interest

Authors declare there is no conflict of interest.

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