

Three Cases of Cutis Marmorata Telangiectatica Congenita

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We report 3 cases of unusual cutis marmorata telangiectatica congenita (CMTC). The ages of the first two cases of CMTC were premature babies, born at intrauterine pregnancy of 31 weeks and 34 weeks; the other patient was a 45-year old female. Compared to the age of other cases reported in the literature, our patients' ages are unusual. It is not surprising that the first two cases of premature babies had typical signs because CMTC is a congenital disorder, usually seen at birth, but the fact that the skin lesion of CMTC appeared already in the early third trimester may provide some clues to the pathogenesis of CMTC. The third case seems to be very rare because most reported cases show that the skin lesion usually disappear gradually over a period of months to years. The site and size of the lesion in the last case was told to have been fixed for 45 years without any change.

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CMTC is a rare vascular anomaly characterized by lasting discoloration of the skin in a reticulate pattern. Since the first report in 1922 by Van Lohuizen¹, more than a hundred cases have been reported in the literature. The skin lesions of a child with CMTC included livedo reticularis, telangiectases and superficial ulceration. The CMTC is also associated with congenital anomalies such as congenital glaucoma and vascular abnormality². But CMTC can exist without any congenital anomaly like our cases. We report three unusual cases of CMTC in point of their ages. The first two cases occurred in premature babies and the last case in a 45-year-old woman.

CASE REPORT

Case 1

A premature baby, born at the IUP (intrauterine pregnancy) of 31 weeks was referred to the department

of dermatology for skin examination. The patient was male, who was born by means of Cesarean section due to placenta previa totalis. Apgar score was 7 at 1 minute and 8 at 5 minute; birth weight was 1.59 kg. From the moment of birth, he had reticulated blue violet colored patches all over his body, which did not disappear by the changes in temperature (Fig. 1). He did not have any other systemic nor dermatological disorders. He did not have any extraordinary family history. His mother was 35 years old and her gestational history was 1-0-5-1. During the midtrimester (IUP 20 weeks), the patient's mother was said to have received medication for common cold symptoms from the department of family medicine. She received oral medication for 2 days consisting of acetaminophen, piroxycam- β -cyclodextrin, chlorpheniramine and intramuscular injection of amoxicillin. Also, she received oral medication of aspirin and dydrogesterone from her obstetrician for vaginal bleeding. From the patient's laboratory examinations, X-ray films of the chest and skull, cranial CT, EKG, metabolic and chromosomal study, amino acid analysis, VDRL and ANA were negative findings or within normal limit.

He was cradled in the incubator and cared by pediatricians. From the clinical manifestation and histological examination (Fig. 4), he was diagnosed with CMTC and was observed, but the alteration of the

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Fig. 1. Case 1; widespread blue-violet colored, reticulated patches over the entire body surface.

Fig. 2. Case 2; widespread blue-violet colored, reticulated patches over the entire body surface.

Fig. 3. Case 3; blue-violet colored, reticulated patch on left knee area.

Fig. 4. Case 1; Histopathology of the affected skin showing dilated vessels in the dermis. (H&E, $\times 200$).

lesion was minimal. The patient was seen in a follow-up visit 4 months after the discharge from hospital. There was some fading of the lesion, but the lesion was not completely cleared.

Case 2.

A premature baby, born at IUP of 33⁶ weeks was

referred to the department of dermatology for skin examination. The patient was male, who was born by Cesarean section due to pregnancy induced hypertension. Apgar score was 7 at 1 minute, 8 at 5 minute, birth weight was 1.26 kg. From the moment of birth, he had reticulated blue-violet colored patches all over his body, which did not disappear by the changes

in temperature (Fig. 2). He did not have any other systemic nor dermatologic symptoms. His mother was 28 years old and her gestational history was 0-0-1-0, and he did not have any extraordinary family history nor past history. Also his mother did not have any medication history during pregnancy.

Laboratory investigations including complete blood count, VDRL and antinuclear antibody were negative or within normal limit. From clinical manifestation, he was diagnosed with CMTC and was observed, but the alteration of the lesion was minimal. 3 months after discharge, on his follow-up visit, there were slight improvements of the lesions, but had not completely cleared.

Case 3.

Forty-five year old female patient with reticulated blue-violet patches on the medial side of left knee visited our office (Fig. 3). She had history of atopic dermatitis, and otherwise nothing extraordinary in her past history. She said that the lesion existed from when she was very young and that she visited for the cosmetic purpose. From clinical sign and histological examination, she was diagnosed as CMTC.

We recommended laser therapy, but the patient refused it. On her follow up visit 2 weeks later, there were no apparent changes in the lesion.

DISCUSSION

In 1922, van Lohuizen described a child with unusual skin lesions that resembled livedo reticularis, accompanied by telangiectases and superficial ulcerations¹. The patient's clinical course was characterized by steady improvement in the appearance of the lesions over an 18-month period. Since then, there have been more than one hundred cases of CMTC reported in the literatures. They have been reported under various names, such as generalized congenital phlebectasis³, naevus vascularis reticularis⁴, congenital livedo reticularis⁵ and van Lohuizen's syndrome⁶. But now, CMTC is used. Lesions are usually on the trunk or extremities, may be localized or generalized, and are often unilateral. The course is variable, but the majority of lesions fade by adolescence. The erythema may be heightened by cooling, physical activity, and crying. The reticulated bands may contain focal areas of atrophy and can result in a limb circumference discrepancy. Ulceration and secondary

infection are frequent complications in severely affected patients⁷. Our cases showed typical reticulated blue-violet patches on the whole body in the first two cases and localized reticulated patch in the last case. But there was no atrophy, ulceration and infection. Especially in the third case, the skin lesions remained unchanged for 45 years. The first case of CMTC associated with congenital anomalies including Sturge-Weber syndrome and patent ductus arteriosus was described by Petrozzi *et al.* in 1970⁸. Associated malformations have been reported in 22% to 68% of patients^{2,9}, but our cases were not associated with other congenital anomalies. Weilepp⁷ and Picascia⁹ noted that CMTC is a relatively benign condition and many of the so-called defects reported in association with this condition, for example, syndactyly of toes, high-arched palate, cleft palate, short stature, and triangular face, might be coincidental expressions of genetic traits. Furthermore, cases of CMTC as a solitary abnormality tend to be underreported because this vascular lesion is well known only to dermatologists. In addition to their notes, some doctors because of its relatively short duration, natural healing tendency and even ignorance may overlook CMTC. In reality, the third patient said that some doctors had ignored her skin sign. In that point of view, the rate of congenital anomalies associated with CMTC may be even lower than that from the literatures.

Histopathologic examinations were done in the first and the last cases. The findings showed vessels that were increased in number and diameter in the dermis. Although the pathologic finding of CMTC is said to be non-specific, most cases of CMTC show similar findings such as dilated capillaries and veins, sometimes expanded to form capillary and venous lakes, throughout the dermis and in subcutaneous tissues¹⁰.

The pathogenesis of CMTC is still unknown, while there are some hypotheses such as genetic basis with an autosomal dominant inheritance^{11,12} and the suggestion that CMTC may be caused by some factors that act during intrauterine growth, at a time crucial for proper vascular development². In most of the reported cases, the skin lesions were found at birth or shortly thereafter, as the nomenclature of CMTC means congenital disease. On the contrary, our first two cases were found in premature babies who were delivered through Cesarean section because they had been in serious obstetric problems. In some cases, the skin lesion did not develop until 3 months after birth, or even 2 years

after birth^{13, 14, 15, 16}. The fact that the onset of the disease is variable may give a clue to the new pathogenesis of CMTc which is different from already postulated. However, it needs more cases like our unusual CMTcs to support such a pathogenesis.

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