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# CUTIS MARMORATA TELANGIECTATICA CONGENITA IN A CHILD - A RARE CASE REPORT

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**ABSTRACT**
Cutis marmorata telangiectatica congenita (CMTC) is an uncommon, congenital, vascular malformation characterized by asymmetrical, fixed reticulate vascular network typically involving the lower limbs, telangiectasia, phlebectasia and occasionally ulceration or atrophy of the affected skin. We report a case of cutis marmorata telangiectatica congenita in a 2 months old male child who presented with complaints of bluish red skin lesions over the limbs, abdomen and both the forearms since birth.

## INTRODUCTION

Cutis marmorata telangiectatica congenita (Syn: Reticulate vascular naevus or Van Lohuizen syndrome) is a rare disorder first described by Cato Van Lohuizen[1] in 1922 with about 300 cases described in the literature to date[2]. The etiology is unknown but it appears to represent a developmental ectasia involving both capillaries and veins. It is a sporadic disorder but may also be transmitted as an autosomal dominant inheritance with variable penetrance. It affects both sexes in equal numbers and is found most often in infants but may also affect adults.

## CASE REPORT

A 2 months old male child born out of consanguineous marriage was brought to our OPD with complaints of bluish red skin lesions present over the both legs, abdomen and both the forearms since birth. No history of darkening of skin lesions on exposure to cold or improvement on rewarming. History from the mother revealed that the child was delivered by full term, normal delivery with birth weight of 2.5kg. Antenatal and perinatal period was uneventful. No history of seizures, feeding difficulties, nasal regurgitation, and cyanotic spells was present. No history of similar complaints in the family.

Differential diagnosis of cutis marmorata telangiectatica congenita, livedo racemosa associated with congenital syphilis, neonatal lupus erythematosus and vascular malformations were considered.

Dermatological examination revealed reticulated reddish blue patches in a net like pattern with telangiectasia seen over the extensor aspect of both the lower limbs (Figure 1), abdomen (Figure 2) and extensor aspect of right forearm (Figure 3) sparing the face. . There was no an obvious limb-length discrepancy, atrophy or ulceration over the vascular areas. The baby has normal facies with a head circumference of 40cm. Palms, soles, scalp, nail and oral mucosa were normal. Systemic examination (including neurological, cardiology and ophthalmology) done was normal.

Complete blood count and coagulation profile done was normal. Skeletal survey and MRI revealed no abnormalities. Serological tests like RPR, TPHA, ANA, and Anti-Ro antibodies done were within normal limits.

**Figure 1. Clinical photograph showing reticulated bluish red patches with telangiectasia over the extensor aspect of both the lower limbs.**

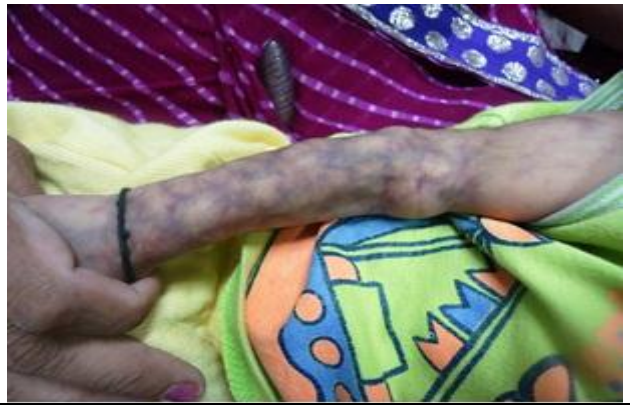


**Figure 2. Clinical photograph showing reticulated bluish red patches with telangiectasia over the abdomen**

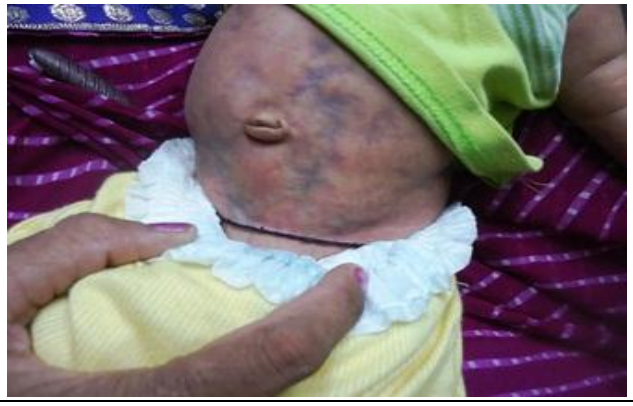
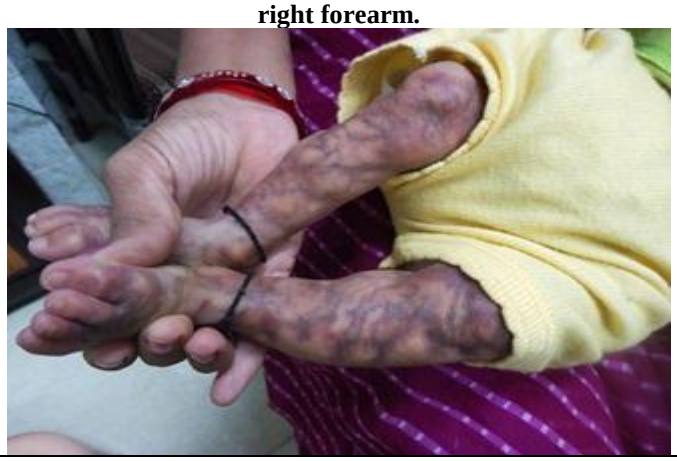


**Figure 3. Clinical photograph showing reticulated bluish red patches with telangiectasia over the extensor aspect of right forearm.**



## DISCUSSION

CMTC is a rare congenital disorder characterized by red-purple vascular network in the skin. Lesions are usually asymmetrical, localized or generalized typically involving the lower limbs less often involving face, trunk and upper extremities. When located on the trunk, the lesions of CMTC tend to show mosaic distribution in streaks with a midline demarcation seen across the abdomen. [3] The reticulated pattern is persistent, fixed and enhanced by cold, crying and exercise. The colour is more vivid and darker and it does not disappear after warming (unlike physiological cutis marmorata). The skin may be atrophic or even ulcerated in the vascular areas. They may manifest as reduced girth of the affected limb and rarely bony hypoplasia. There are numerous other congenital anomalies reported in association with CMTC with a prevalence of 20-70%. The most common defects are aplasia cutis, developmental delay and cleft palate. The common associations are macrocephaly (macrocephaly cutis marmorata telangiectatica congenita syndrome), vascular anomalies (capillary and cavernous hemangiomas, Sturge-Weber syndrome, Klippel-Trenaunay syndrome, Adams Oliver syndrome, nevus flammeus) [4,5], hemiatrophy, glaucoma and retinal detachment[6], neurological abnormalities, hypothyroidism, scoliosis and anogenital abnormalities.

The differential diagnosis of CMTC are physiological cutis marmorata, neonatal lupus erythematosus (where head is affected and skin changes are bilaterally symmetrical) [7,8], livedo racemosa, vascular and lymphatic malformations like port-wine stain and Klippel-Trenaunay syndrome, metabolic disorders like homocystinuria and rarer conditions like Divry Van Bogaert syndrome [9], Cornelia de Lange syndrome, Adams-Oliver syndrome and livedo reticularis associated with collagen vascular disorders. Histologically, it is characterized by dilatations of capillaries and venules in the dermis, a proliferation of vascular channel. The patient rarely requires treatment as it is self limiting and many children improves in the first 2 years of life. For those with persistent reticulate erythema, pulsed dye laser may be

used to lighten the affected patches. This case is reported because of its rarity.

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**CONFLICT OF INTEREST**

The authors declare that they have no conflict of interest.

**STATEMENT OF HUMAN AND ANIMAL RIGHTS**

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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