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Case Report

An Interesting Case of Persistence Reticulate Erythema… Cutis Marmorata Telangiectatica Congenita

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Abstract

Cutis marmorata telangiectatica congenita (CMTC) is a benign angiomatous condition usually seen at birth as dilation of superficial capillaries and veins. Its characteristic feature is red or purple reticulated anastomosed macules having localized or generalized asymmetrical appearance. These lesions may also be associated with ulceration or atrophy. It is a rare disease with unknown etiology. However, prognosis is generally good.

Here, we are presenting a case of male term baby born at 40 week gestation with APGAR score 5/1 and 9/5 and no associated abnormalities. The management of such a case requires involvement of a pediatrician, dermatologist and psychologists.

Keywords: *Cutis Marmorata Telangiectatica Congenital; Diagnosis; Prognosis*

Introduction

Cutis marmorata telangiectatica congenita (CMTC) is a rare vascular skin condition, recognized by Von Lohuizen in 1922. It is also known as VON LOHUIZEN disease [1]. It is a benign dermatological condition seen at birth with dilation of superficial capillaries and veins, characterized by red or purple reticulated anastomosed macules which are asymmetrical either localized or generalized [2]. Although described as rare, the incidence of CMTC is probably underestimated as it is based on clinical findings [3].

Case Report

A 26 years old mother, gravida 3 para 1+1 gave birth to a male baby at 40 week gestation. The pregnancy term was uneventful and baby delivered vaginally. The newborn was eutrophic, weighing 3.2 kg and head circumference was within normal limits (34 cm). Immediately after birth, baby was shifted to nursery where baby was vitally stable and systemic examination was unremarkable.

On skin examination, multiple bruises appearing as a spider's web noted. These skin lesions were seen on right side of body including the entire trunk, arm and leg sparing face. It did not disappear during the warming of the baby. A clear demarcation between normal and involved skin was noted as shown in figure 1-3.

The baby had no facial dysmorphic features. To rule out any other conditions cranial ultrasound, abdominal doppler studies, thyroid function and karyotyping were performed. All reported normal. Hence on the basis clinical findings, investigations performed and exclusion of any other disease, diagnosis of Cutis Marmorata Telangiectatica Congenita (CMTC) was established. The baby had no findings on ophthalmological examination and was discharged on the 4th day of life with a scheduled follow-up appointment.

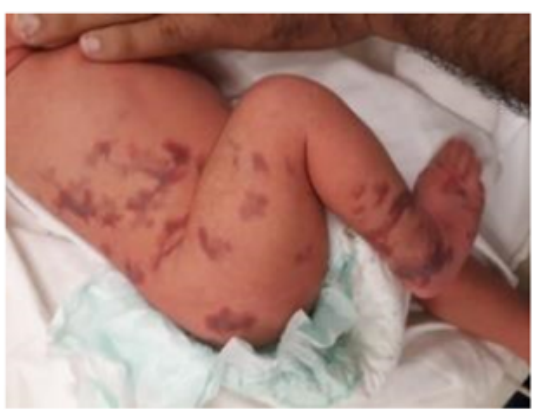

Figure 1

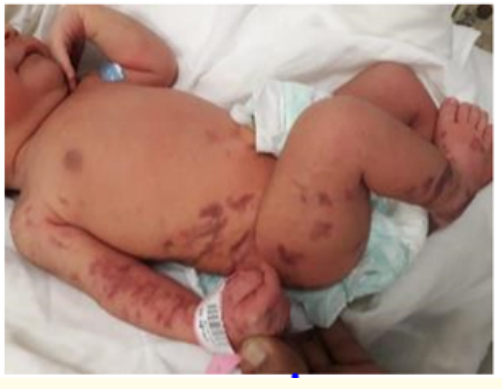

Figure 2

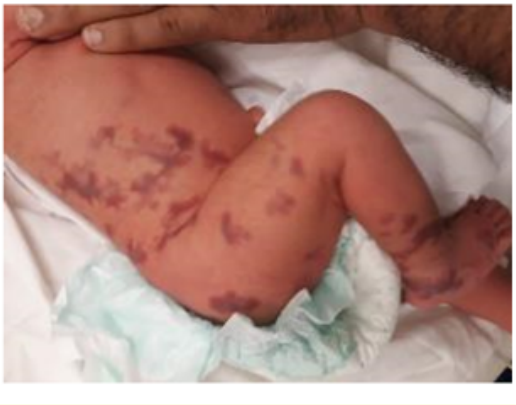

Figure 3

Outcome and follow-up

Regular follow-up was arranged to allow early detection of other features of the disease. Purpuric reticulate lesions would improve gradually, reducing pigmentation extension. At six month follow-up it was observed that lesions were mainly confined to the lower limbs. The cephalic perimeter remained stable at the 50th percentile.

Discussion and Conclusion

Cutis marmorata telangiectasia congenita (CMTC) is a rare congenital vascular anomaly presenting with localized or generalized persistent telangiectasia with characteristic reticulate erythema pattern, maybe associated with skin atrophy and ulceration [4]. About 300 cases of this disease are reported in literature [1]. 95% CMTC presents with skin changes at birth. However, lesions may also present later. Equal sex distribution was observed [2].

Though etiology and pathogenesis of CMTC is unknown, the typical segmental distribution with midline separation suggests that it may be caused by genetic mosaicism [5]. Nevertheless, other studies advocate an autosomal dominant inheritance pattern with incomplete penetrance [6]. All reported cases were sporadic. There were no familial cases recognized, as some theories propose that CMTC may have a genetic support [7,8]. Literature reports only one case of CMTC associated to a macrocephaly by Stoll in 2003 with a novo translocation t(2;17) (p11;p13) [9].

Physiological Cutis Marmorata can be considered as differential diagnoses of CMTC, resulting due to low skin temperatures and disappears upon warming [10]. The skin anomalies are often characteristic to make the diagnosis [11].

Occurrence of vascular complications in addition to the vascular skin changes may confuse it with generalized vasculopathy [12]. The prognosis is usually reported well, with slight concomitant anomalies. Literature suggests that in 50% of the babies, the lesions disappear during the first 2 years of life. However, scarring lesions will remain unchanged throughout the patient's life [13].

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