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Cutis Marmorata Telangiectatica Congenitale with Neonatal Revelation

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Abstract

Cutis marmorata telangiectatica congenita (CMTC) is a rare congenital disorder of un known etiology. It ischaracterized by a reticulated erythema, called cutis marmorata, as well as phlebectasia and telangiectasia. Unlike the benign cutis marmorata, it does not disappear with warming. The CMTC may have extra coetaneous manifestations. Diagnostic criteria have been suggested to facilitate the diagnosis of congenital cutis marmorata telangiectatica, but are not yet validated. Prognosis is variable. A routine multi disciple unary follow-up is suggested in the first years of life to monitor the appearance of new abnormalities.

Keywords: Cutis marmorata telangiectati cacongenitale, neonatal, vacuities

Introduction

Cutis marmoreal elangiectatica congenita (CMTC) is a rare congenital vascular disease, described for the first time in 1922 by Van Louise [1]. Skin lesions are observed from birth or shortly thereafter, and may increase during the first weeks of life. CMTC is manifested by the presence of a blue-violet vascular network in the skin, reticulated and localized or generalized, often asymmetrical. The cutaneoussigns range from fine and diffuse capillary abnormal ities without atrophy to wide, violet and ulcerate reticulate bands.

The cutaneouslesionsappearmostoften on the legs, sometimes on the arms and the trunk, and rarely touch the face and the scalp [2].

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Observation

This is a newborn male; from a non-consanguine nous marriage; a 20-year-old mother with no notable antecedents; pregnancy was normal carried out with a positive infectious history on prolonged membrane rupture lasting 20 hours; delivery was high with a good adaptation to extra uterine life. He was admitted to the Intensive Care Unit for Infectious Disease, where the clinical examination found a euro phi new born, Weight = 3100g; Size = 51cm; Pc = 33cm; without facial dimorphism or clinically detectable malformation; hem dynamically and respiratory stable. The coetaneous examination revealed a blue-violet vascular network with a cross-linked appearance located opposite the right hypo chondriumwith extension towards the back without there capillary malformation or hypotrophy of the ipsilichemi-body. The infectious balance was negative and the extension assessment did not reveal any other localization, namely normal funds; normal brain imaging and liver ultrasound. During the follow-up; the infant had a good psycho motor development at the age of 3 months without extension of the cut an emulsions.

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Discussion

The cutis marmorata telangiectatica congenitais in frequent. About 300 cases have been reported so far. Both sexes are equally affected [3]. CMTC lesions are usually present birth as reported; with some lesions develop in gloater between 3 months and 2 years of age [4]. The pathogenesis of CMTC is not yet known, although several factors were in criminated according to the literature [5]. It ischaracterized by a dilation of the veins and capillaries of the coetaneous and subcutaneous tissue, anatomized in networks giving a cross-linked (or marbled) appearance. Bluish venous vessels may be visible by transparency, skin ulcers and atrophic areas may be noted in places. CMTC can be diffuse all over the body including the scalp or limited to a part of the body as our case [10]. Extra coetaneous manifestations have been reported in 20 to 80% of patients with CMTC [6]. These rates have been criticized for allege overlap with other concurrent genetic disorders [7]. The mostcomm onextracutan eousfindingis body asymmetry, particularly of limbs, as reported by the different Devilries studies (43%), Keenest (33%) and Per et al. (68%) [8; 6]. Glaucoma has also been reported, although rare and often associated with other vascular disorders such as nevi [1]. However; none of these extraskin manifestations were found in our patient. The association of a CMTC with other syndromes is possible; itisdescribedin 20-25% of reported cases of Adams-Oliver syndrome [9].

The diagnosis is essentially clinical; must be accurate and early in view of the risk of life-threatening complications and neurological abnormalities throughout life. The association with extra-skin manifestations, as previously indicated, makes the differential diagnoses quite broad, such as Klippel-Trenaunay syndrome, Surge-Weber syndrome, and Bockenheimer's syndrome; some facial angioma capillary malformations, and macro cephalic -

CMTC. The persistence of cut an emulsions under local warmingmakesit possible to differentiate CMTC from physiological cutis marmorata [11]. In general, no treatment is necessary. The results of laser therapy in patients with persistent CMTC differ according to the studies [12].

An annual multi disciplinary follow-up is recommended for at least 3 years. Long-term follow-up is rarely reported [8].

The prognosis of CMTC is generally good. About 50% of patients have sporting eousresolution of skin manifestations that usually occur before the age of 2 years. Factors that predict the resolution of lesion stilled to be identified [12].

Conclusion

Cutis marmoreal elangiectatica congenital is a rare condition of un know etiology. It ischaracterized by localized or generalized reticulate derythema; associated or not with other coetaneous manifestations. Diagnostic criteria have been suggested to facilitate diagnosis, but are not yet validated. A multi disciple in aryl follow-up during the first years of life is necessary.

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Legends Figure

Figure 1: image showing the CMTC located next to the right hypo chondrium with extension to the back in our patient

