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What is your diagnosis?

Tanınız Nedir?

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Congenital reticulate erythema in a newborn

A female newborn was applied with brownish marbled and purple reticulated skin lesions involving patients' the right leg (Figure 1). She was spontaneously delivered, 2600 gram with Apgar score was 9-9 at first and fifth minutes of life respectively at 38 weeks. Family medical history was noncontributory. She was otherwise healthy. Neurological and ophtalmological examination and routine labaratory investigations were normal. In her dermatological examination, unilateral marmorated brown-purple skin lesions with atrophy within the affected area and ulceration were seen. Lesion was unresponsive to local warming. There wasn't any other vascular anomalies or asymmetry of limbs.

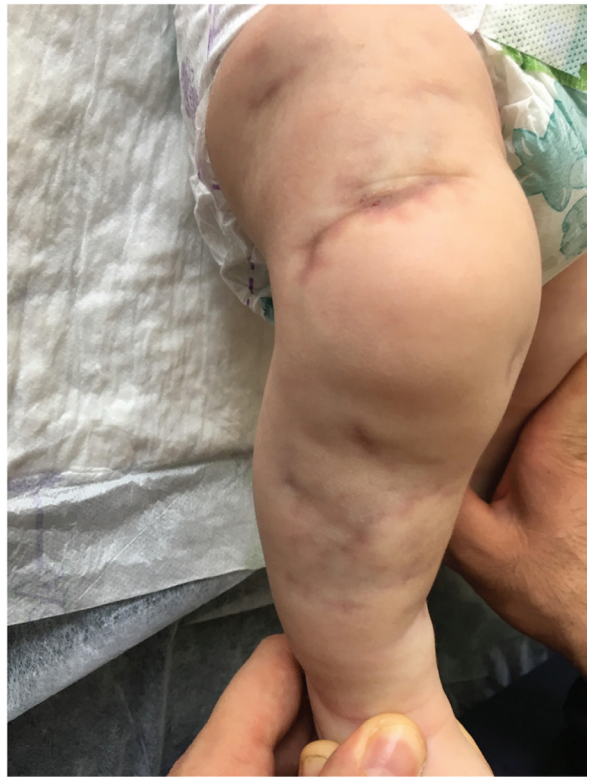

Figure 1. Brownish marbled and purple reticulated skin lesions involving patients' the right leg, and atrophy within the affected area

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Diagnosis: Cutis marmorata telangiectatica congenita

Clinical characteristics of patient prompted the diagnosis of cutis marmorata telangiectatica congenita (CMTC). CMTC is an uncommon sporadic congenital vascular anomaly that usually presents at birth. Various anomalies being associated with CMTC. The rate of anomalies reported with CMTC varies between 18.8% and 70% in medical literature[1-4]. Limb asymmetry (hypoplasia or hyperplasia) and vascular anomalies are most commonly reported anomalies, followed by neurological disorders, ocular malformations and syndactyly[5]. Kienast and Hoeger[1] reported body asymmetry 33% in 27 CMTC patients which they were followed up prospectively. Since there is no histopathological diagnosis of disease, Kienast and Hoeger[1] suggested diagnostic criterias (Table 1). Although it needs to be evaluated in future prospective studies, it seems useful and practical and also helpful to distinguish CMTC from the other vascular anomalies.

Although it is a benign disease which is self limiting, a complete examination and multidisciplinary follow-up should be performed in order to evaluate related possible anomalies. CMTC should distinguish from other conditions that may mimic CMTC like Klippel-Trenaunay syndrome, Sturge-Weber syndrome, Adam-Oliver syndrome, Bockenheimer's disease. Other differantial diagnosis includes physiological cutis marmorata and reticular hemangioma sydrome. Physiological cutis marmorata shows a similar pattern with CMTC but disappears with local heating. In Klippel-Trenaunay syndrome, venous varicosities, port wine stains and soft-tissue hypertrophy can be seen. Sturge-Weber syndrome can be present by facial port wine stain, mental retardation, seizures, cerebral malformations, glaucoma, and cerebral tumors. In Adams-Oliver syndrome limb defects, cardiac malformations and aplasia cutis can be seen[6]. Bodenheimer disease (diffuse phlebectasia) characterized with diffuse large and mostly painful venous ectasias which onset at childhood and progressed. Reticular hemangioma syndrome characterized with anogenital-urinary-sacral anomalies, and sometimes cardiac failure[7].

CMTC usually doesn't require treatment and prognosis is good. More than half of the patients result in fading of erythema and marmoration during the first two years of life[8]. Vasodilators, aspirin, pentoxifylline, avoidance of cold, psoralen and ultraviolet A and laser therapy were reported treatments that have shown varying results[9]. If ulceration is present, treating infected areas and using occlusive dressing are recommended. Annual follow-up is neccesary for screening associated anomalies for at least 3 years.

Table 1. Kienast's suggested diagnostic criteria for CMTC

Major criteria
- Congenital reticulate (marmorated) erythema
- Absence of venectasia
- Unresponsiveness to local warming
Minor criteria
- Fading erythema within 2 years
- Port wine stain outside the area affected by CMTC
- Telangiectasia within the affected area
- Skin ulceration within the affected area
- Atrophy within the affected area
*Presence of all three major criteria and at least two or more minor criteria
CMTC: Cutis marmorata telangiectatica congenita

Ethics

Informed Consent: Informed consent taken from patients' legally parents.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: E.Ö., Design: E.Ö., Data Collection or Processing: T.F.G., Analysis or Interpretation: İ.K.A., Literature Search: E.Ö., Writing: E.Ö.

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