PAEDIATRIC DERMATOLOGY

RARE CASE OF CUTIS MARMORATA TELANGIECTASIA CONGENITA

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Background: Cutis marmorata telangiectatica congenita (CMTC), first described by Dutch paediatrician Van Lohuizen in 1922 is an exceedingly rare congenital condition, with about 300 cases described in the literature to date. The disorder presents with a localised, segmental, or generalised persistent reticular mottling and closely resembles cutis marmorata, a common benign response observed in infants, which resolves with warming of the skin surface. In contrast, CMTC lesions don't disappear with rewarming and occasionally present with ulceration and atrophy of the involved skin.
Many associated exocutaneous malformations have been reported including asymmetry of the involved limbs, ocular abnormalities, growth and developmental delays, neurological abnormalities, as well as additional vascular anomalies including angiokeratomas, hemangiomas and port-wine stains. Cutaneous lesions carry a good prognosis and tend to improve with age.

Observation: A full- term, 2-day-old female with a congenital non-tender reticular patch, that did not disappear with local warming was referred to our department for consultation. The family history as well as the antenatal course and delivery were unremarkable. On examination, a marbled bluish to deep purple lesion appeared to extend over the right side of the right side of her body, face and scalp, and there was presence of atrophy of the involved skin, along with ulceration above the right lateral malleolus. Head and limbs circumferences were within normal range. The patient was checked by the ophthalmology and neurology department to screen for associated anomalies, which were not detected. On the grounds of medical history and clinical presentation, the diagnosis of CMTC was established.

Key message: The diagnosis of CMTC is made on clinical grounds and skin biopsy is not required if the clinical diagnosis is definite. A thorough screening for associated anomalies as well as annual controls of skin changes and psychomotor development of the patients should be performed.