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CUTIS MARMORATA TELANGIECTATICA CONGENITA*

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Cutis marmorata telangiectatica congenita (CMTC) is a rare and clinically distinctive cutaneous vascular anomaly present at birth, which frequently improves with age, and may be associated with other abnormalities in at least 50% of the patients[1,2]. The syndrome was first described by van Lohuizen[3] in 1922 and is typified by five features: congenital persistent cutis marmorata, phlebectasia, telangiectasia, ulceration, and slow clinical improvement[4]. There have been scattered reports of CMTC in the literature[2-11] under the synonyms of congenital generalized phlebectasia[12-17], nevus vascularis reticularis[1,2], and congenital livedo[18]. The cause of CMTC is still quite unknown[1]. There is no apparent genetic inheritance of the anomaly[1,9,11].

In this paper we would like to report a case of CMTC in a 3-year-old boy; this is, to the best of our knowledge, the first such case to be reported from Turkey.

Case Report

A 3-year-old boy was admitted to the Pediatrics Department of Karadeniz University Faculty of Medicine because of his generalized mottled appearance. He had looked normal at the time of birth, but the next day the family had noted a generalized mottled red appearance which was more prominent on the right arm. No parental consanguinity was recorded. His parents and a five-year-old sibling were in good health.

On physical examination the patient had a generalized mottled red appearance. The vascular markings on his right arm were more prominent than on the other

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parts of the body. There was no ulceration or scarring on the skin (Figure 1). The results of the rest of the physical and neurologic examination were normal. The results of all routine laboratory tests were within normal limits. A punch biopsy (5 mm in diameter) of the skin of the right arm revealed normal findings. The patient's ailment was diagnosed as generalized CMTC with localized distribution.

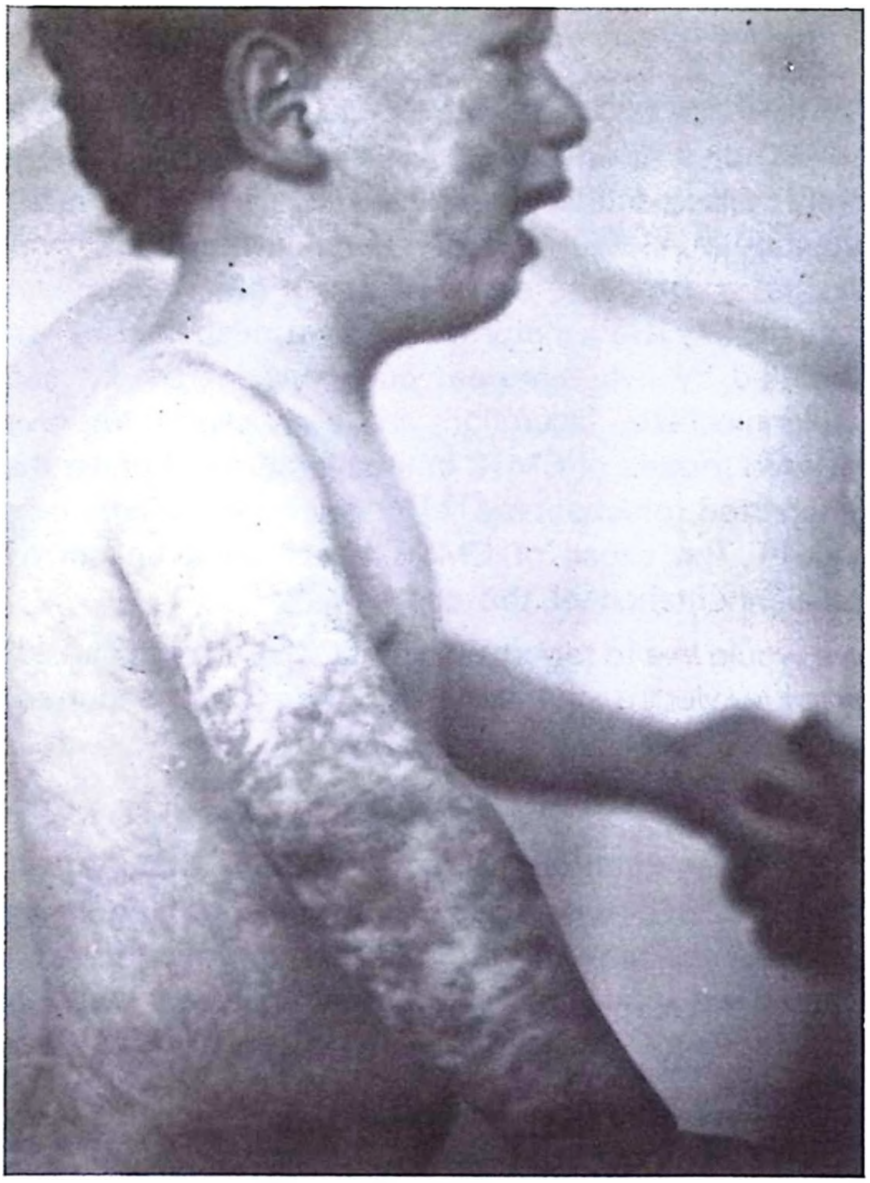

Figure 1
The vascular markings on the right arm of the patient

Discussion

Cutis marmorata telangiectatica congenita is a distinct clinical entity characterized by prominent veins and capillaries in the superficial skin, giving a mottled bluish appearance. It occurs in either a localized (segmental) or generalized distribution[1,2]. As reported by van Lohuizen[3], the major features include

congenital persistent cutis marmorata, phlebectasia, telangiectasia, ulceration with crusts, and a benign course with gradual improvement.

Way et al[9], in a review of the world literature, found a female preponderance of 24:38, and an earlier review of the English literature had revealed only seven original case reports of classic CMTC in males[6,9,10,18]. A 1978 study, however, reported that 50% of the patients were boys, the vast majority of the male patients having a localized distribution[2]. Our male patient with localized distribution conforms to that observation.

As previously reported, CMTC involves, by order of frequency, the extremities, trunk, face and scalp[1,2,9]. Localization on the palms, soles and mucous membranes is infrequent. Associated anomalies occur in about 50 percent of the cases[1,9] and include subcutaneous atrophy[9,17], deep ulceration[6], hemiatrophy[8], hemihypertrophy[9], dystropic teeth[12], glaucoma[7], patent ductus arteriosus[7], mental retardation[7], Sturge-Weber syndrome[7], macrocephaly[10], nevus flammeus[7,10], varicosities[9,11], capillary and cavernous hemangiomas with or without the Klippel-Trenaunay syndrome[9], syndactyly[9] and delayed psychomotor development[9]. Physical growth and development have been normal in most patients and there have been no instances reported of malignant degeneration[9].

Most of the cases in the literature were showing definite signs of fading within the first years[1,2]. It has been theorized that this may be due to the normal maturation process with thickening of the epidermis and dermis[9]. However, there have been reports of persistence into adulthood[7].

There has been no evidence in any previous reports of a genetic basis for this condition[1,9,11]. However, Kurczynski[19] recently reported a child with CMTC whose father and paternal grandmother had also had the disorder, although it had faded in adult life. This suggests autosomal dominant inheritance, but further studies will be necessary to establish the genetic basis. In this case there was no similar problem in any other members of the family.

In differential diagnosis, physiologic cutis marmorata needs to be considered. There is no available treatment for CMTC. In our own case there were no associated abnormalities. The patient was not given any treatment.

There are no specific microscopic findings in CMTC[1,2]. Van Lohuizen's original patient had enlarged dilated veins, capillary and venous lakes, and large capillaries in all layers of dermis and subcutis[1-3]. Others have found a similar pattern, without inflammation or other change in the skin and with no abnormality on special staining[15-17]. No vascular abnormalities were found by Petrozzi et al[7], nor did we find any in our patient.

Summary

The case of a 3-year-old boy with cutis marmorata telangiectatica congenita is presented. This is believed to be the first report of its kind from Turkey.

REFERENCES

1. Jacobs AH. Vascular nevi. *Pediatr Clin North Am* 30:467, 1983.
2. South DA, Jacobs AH. Cutis marmorata telangiectatica congenita (congenital generalized phlebectasia). *J Pediatr* 93:944, 1978.
3. van Lohuizan CHJ. Über eine seltene angeborene Hautanomalie (Cutis marmorata telangiectatica congenita). *Acta Dermat-venereol* 3:202, 1922.
4. Moyer DG. Cutis marmorata telangiectatica congenita. *Arch Dermatol* 93:583, 1966.
5. Neumark S. Zur Kenntnis der Livedo telangiectatica congenita generalisata. *Acta Dermat-venereol* 19:316, 1938.
6. Pierini LE, Grinspan D. Cutis marmorata telangiectasia congenita. *Arch Argentinos Dermatol* 5:295, 1955.
7. Petrozzi JW, Rahn EK, Mofenson H, et al. Cutis marmorata telangiectatica congenita. *Arch Derm* (Chicago) 101:74, 1970.
8. Fitzsimmons JS, Starks M. Cutis marmorata telangiectatica congenita or congenital generalized phlebectasia. *Arch Dis Child* 45:724, 1970.
9. Way BH, Hermann J, Gilbert EF, et al. Cutis marmorata telangiectatica congenita. *J Cutan Pathol* 1:10, 1974.
10. Stephan MJ, Hall BD, Smith DW, et al. Macrocephaly in association with unusual cutaneous angiomatosis. *J Pediatr* 87:353, 1975.
11. Gellis SS, Feingold M. Cutis marmorata telangiectasia congenita (congenital generalized phlebectasia). *Am J Dis Child* 131:1027, 1977.
12. Senear FE, Caro M. Generalized telangiectasis. *Arch Dermatol* 23:181, 1931.
13. Humphries JM. Generalized congenital phlebectasia. *J Pediatr* 40:486, 1952.
14. Bedell RF, Allison JR. Congenital generalized phlebectasia in a newborn. *Arch Derm* (Chicago) 90:83, 1964.
15. Kantor I, Yep D. Congenital generalized phlebectasia. *Arch Dermatol* 93:774, 1966.
16. Mizrahi AM, Sachs PM. Generalized congenital phlebectasia. *Am J Dis Child* 112:72, 1966.
17. Lynch PJ, Zelickson AS. Congenital phlebectasia. A histopathologic study. *Arch Derm* (Chicago) 95:98, 1967.
18. Champion RH. Livedo reticularis. A review. *Br J Derm* 77:167, 1965.
19. Kurczynski TW. Hereditary cutis marmorata telangiectatica congenita. *Pediatrics* 70:52, 1982.