Cutis Marmorata Telangiectatica Congenita: A Rare Clinical Manifestation of Capillary Hemangioma?

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A Histologic finding of the Cutis marmorata telangiectatica congenita(CMTC) is non-specific, but dilated capillaries and increased number of vessels are often observed. Capillary hemangioma, mostly represented by strawberry nevus, may show various clinical manifestations. We report a case of Cutis marmorata telangiectatica congenita, which may be an unusual clinical presentation of capillary hemangioma. (Ann Dermatol 15(4) 166~168, 2003).

Key Words : Capillary hemangioma, Cutis marmorata telangiectatica congenita

Cutis marmorata telangiectatica congenita(CMTC) is a rare congenital vascular disease. The major clinical features are persistent cutis marmorata, spider nevus-like telangiectasia, phlebectasia, and occasional ulceration. These features show improvement with age or remain persistent[1]. Up to 50% of reported cases have an associated abnormality including hypoplasia or hyperplasia of the affected or unaffected limb, and vascular lesions such as capillary hemangiomas and telangiectasias[2]. However, CMTC associated with capillary hemangioma on the same site is extremely rare. We report a case of CMTC showing histologic features of capillary hemangioma.

CASE REPORT

A 19-day-old baby had blue violet colored reticulated patches, focal depressed plaques on the right leg, and telangiectasia, which did not disappear by changes in temperature(Fig. 1). The length and diameter of both legs were the same. He did not have any other systemic abnormality nor dermatological disorders. Laboratory examinations including chest X-ray, and abdominal ultrasonography showed negative findings or within normal limit. Histopathological examination of focal depressed plaque on the right leg showed dilated capillaries throughout the dermis(Fig. 2A), and vascular lobules separated by fibrous septa in the subcutaneous tissue. The lobules were composed of plump endothelial cells and small vascular channels(Fig. 2B). When the patient was seen 100 days after the first visit, there was some fading of the lesion, but the lesion was not completely cleared(Fig. 3).

DISCUSSION

CMTC is a rare, usually congenital vascular disease. In 1922, Van Lohuizen[3] first described CMTC, and its major clinical features are persistent cutis marmorata, spider nevus-like telangiectasia, phlebectasia, and occasional ulceration. Sites of prediction are extremities, trunk, face, and scalp[1]. The skin lesions show a marked improvement over time, with the greatest improvement occurring during the first and second year of life[4]. Additional anomalies have been frequently reported in association with CMTC. The most commonly associated findings include body asymmetry(usually limb hyperplasia or hypoplasia), vascular anomalies(mostly

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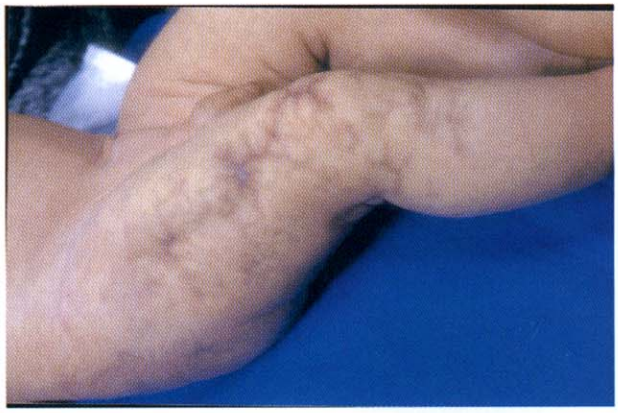

Fig. 1. Erythematous to violet colored reticulated patches with focal atrophy on the right leg.

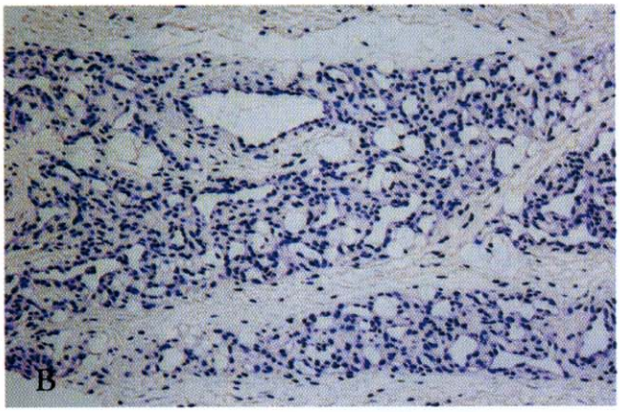

Fig. 2B. Band-like vascular proliferation composed of plump endothelial cells and small vascular channels in the subcutaneous tissue(H & E stain, ×100).

capillary malformation), glaucoma, hypoplasia or aplasia(ranging from transverse limb defects to localized aplasia cutis congenita and cleft palate), and infrequently psychomotor and/or mental retardation.

The histopathologic findings of the cutaneous lesions are non-specific and may show no vascular abnormalities[5,6], but dilated capillaries and veins, vascular fibrosis, and increased number of vessels are most frequently observed. Less common findings include hyperkeratosis, perivascular infiltration, dilated lymphatic veins, and venous thrombosis[7]. Our case showed dilated capillaries throughout the dermis as previous CMTC cases, but the subcutaneous component looked more likely capillary hemangioma: highly cellular vascular lobules composed of small vascular channels and plump endothelial cells. The reticulated pattern of CMTC is believed to represent an abnormal dilatation of capillaries and veins[8].

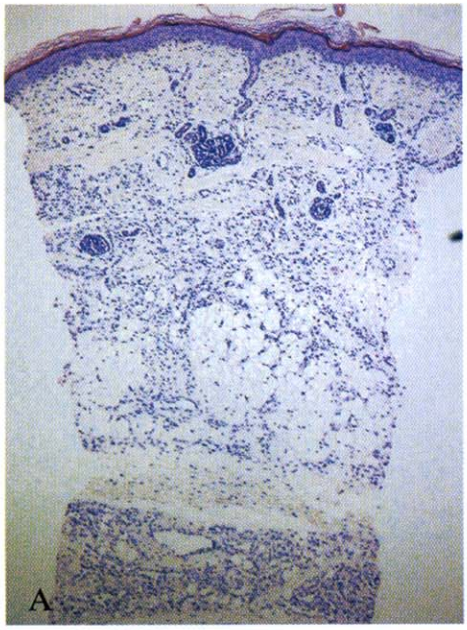

Fig. 2A. Telangiectasia throughout the dermis(H & E stain, ×40).

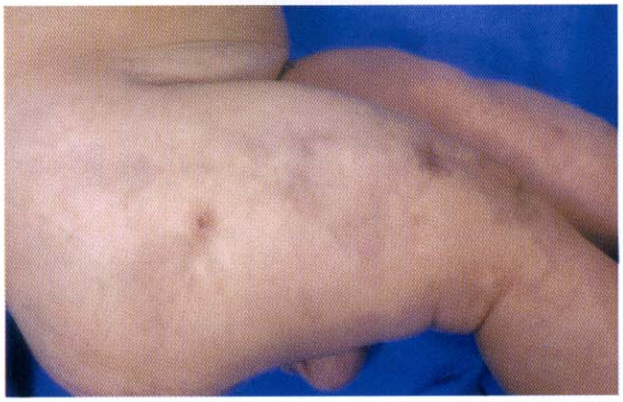

Fig. 3. Fading of the previous lesion is observed.

Clinically, CMTC must be differentiated from other congenital or acquired disorders with reticular lesions, livedo vascular lesions, telangiectases, and/or phlebectases such as cutis marmorata, congenital livedo, angioma serpiginosum, diffuse genuine phlebectasia, generalized essential telangiectasias, Klippel-Trenaunay-Weber syndrome, and Sturge-Weber syndrome[7]. The following clinical features are commonly present and important in the diagnosis of

CMTC; 1. persistent cutis marmorata; 2. spider nevus like telangiectasia; 3. phlebectasia; 4. bilateral involvement, although there is often predominant distribution on one side of the body; 5. early onset of skin lesions, usually present at birth, occasionally within two years of age; 6. improvement of cutis marmorata, especially in the first two years of life; 7. association with other congenital physical anomalies; and 8. dilated capillaries or veins in skin biopsy specimens. The following clinical findings are uncommonly present but helpful in establishing the diagnosis; 1. ulcer; 2. atrophy of cutis or subcutis with thin and transparent skin; 3. capillary hemangioma or venous hemangioma associated with other skin manifestations; and 4. hyperkeratosis or dilated lymphatic vessels on skin biopsy specimens[1]. Our case shows clinical and some pathological features of CMTC, including persistent cutis marmorata, spider nevus-like telangiectasia, early onset of skin lesions, improvement of cutis marmorata, especially in the first two years of life, and dilated capillaries or veins in skin biopsy specimens.

Capillary hemangioma, mostly represented by strawberry nevus, constitutes the most common vascular tumor of infancy. Lesions usually first appear between the third and fifth week of life, increase in size for several months to one year, and then start to regress. Histopathologically, proliferation of endothelial cells are shown, and endothelial cells are large, mitotically active, and aggregated predominantly in solid strands and masses[9]. But, clinically, not all hemangiomas look like strawberries. Martinez-Perez et al.[10] reviewed photographs of over 500 children with hemangiomas, 4% were determined to have an unusual presentation. They were grouped into four morphologic categories; 1. deep, subcutaneous, and/or intramuscular hemangioma with normal overlying skin; 2. macular hemangioma with port-wine stain-like appearance; 3. bossed hemangioma with telangiectasia and peripheral pallor, and; 4. hemangioma with persistent fast flow stimulating arteriovenous malformation[3]. Amitai et al.[11] believed that capillary hemangioma may represent an extension of CMTC because of the high rate of accompanying capillary hemangioma in CMTC and their grossly common pathology. The histopathologic feature of capillary hemangioma in CMTC in our case makes us think that CMTC may be a rare clinical presentation of capillary hemangioma.

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