Congenital Smooth Muscle Hamartoma : a Patchy Follicular Variant

Soon Baek Kwon, M.D., Seok Jong Lee, M.D.,
Do Won Kim, M.D., Jae Bok Jun, M.D.

Department of Dermatology, School of Medicine, Kyungpook National University, Taegu, Korea

Cutaneous smooth muscle hamartomas are benign proliferations of smooth muscle bundles within the dermis. They can be congenital or acquired, and most cases are congenital. Congenital smooth muscle hamartomas (CSMHs) usually manifest at birth as well-circumscribed, frequently hypertrichotic, hyperpigmented or skin-colored patches or plaques on trunk or extremities.

We report a case of CSMH in a 10 year-old girl, who showed a localized skin-colored patch showing prominent follicular papules on the lateral aspect of her right upper arm, which were found at birth. There was no hypertrichosis and the pseudo-Darier sign was negative. This patchy follicular variant is the less common clinical type of the disease.

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Key Words : Smooth muscle hamartoma, Congenital, Follicular variant

Congenital smooth muscle hamartomas (CSMHs) usually appear at birth as skin-colored or slightly hyperpigmented patches or plaques that often contain increased vellus hairs. Histopathologically, it represents a proliferation of bundles of smooth muscle within the dermis[1-4].

We report here a case of CSMH showing a skin-colored patch with marked follicular papules on the right upper arm. There was no hypertrichosis. This patchy follicular variant of CSMH is the less common clinical type of the disease, and differs from those of the previous reports[5-7] in the Korean dermatologic literature.

CASE REPORT

A 10 year-old girl presented with a skin-colored patch on her right upper arm that had been present since birth. Examination revealed a circumscribed, child-palm-sized patch with prominent follicular papules on the lateral aspect of right upper arm. There was no hyperpigmentation or hypertrichosis (Fig. 1). A pseudo-Darier's sign could not be elicited. The patch was asymptomatic and no functional abnormality of the limb was noted. Her medical history was uneventful and there was no family history of similar lesions. Laboratory studies including complete blood cell count, blood chemistry, urinalysis, chest-X-ray and EKG were within normal limits or negative. She had no other developmental anomaly and height and body weight were within normal range of her age group. Clinical differential diagnosis included CSMH, Becker's nevus, congenital nevocytic nevus, and lichen spinulosus. An incisional biopsy was performed. Histopathologically, the epidermis showed mild hyperkeratosis, acanthosis, and slightly elongated rete ridges, but, basal hyperpigmentation was not prominent. Numerous, various sized, bundles of smooth muscle were scattered throughout the dermis, mostly unattached to the hair follicles (Fig. 2A & 2B). Trichrome and actin stains confirmed the presence of smooth-muscle bundles (Fig. 3A & 3B).

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Reprint request to : Seok Jong Lee, M.D., Department of Dermatology, School of Medicine, Kyungpook National University, Taegu, Korea
Tel : (053)420-5821, Fax : (053)426-0770
E-mail : seokjong@knu.ac.kr

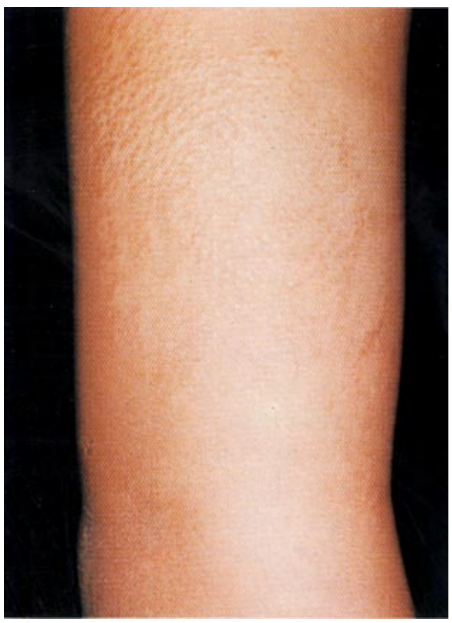

Fig. 1. A child-palm sized, skin-colored patch with marked follicular papules on the lateral aspect of right upper arm.

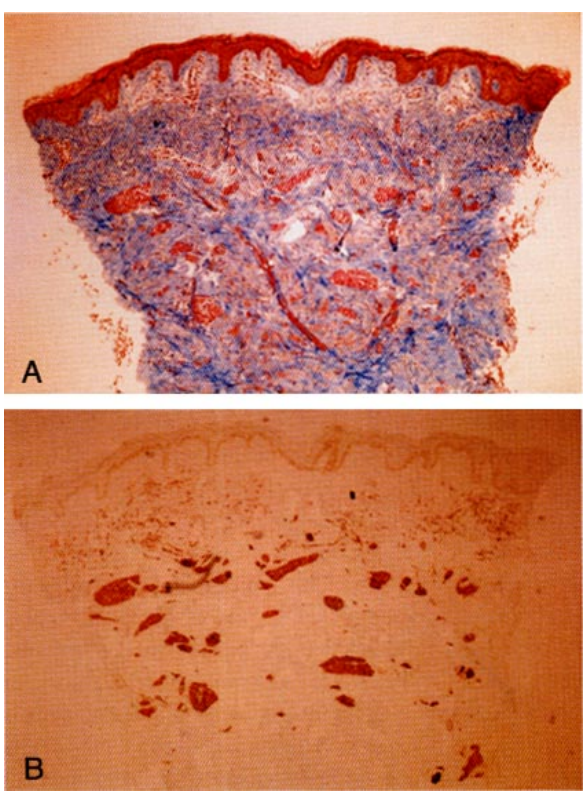

Fig. 3A & 3B. Proliferation of bundles of smooth muscle within the dermis (Masson's trichrome(A) and actin(B) stain, ×40/×40).

Fig. 2A. The epidermis shows mild hyperkeratosis, acanthosis, and slightly elongated rete ridges, but basal hyperpigmentation is not prominent (H&E, ×40).

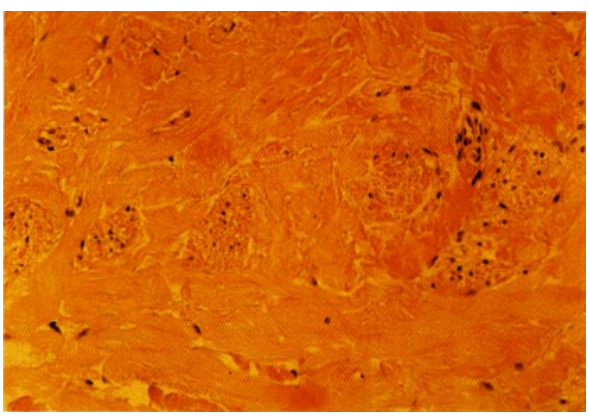

Fig. 2B. Marked proliferation of bundles of smooth muscle within the dermis (H&E, ×100).

DISCUSSION

Cutaneous smooth muscle hamartomas are benign proliferations of smooth muscle bundles within the dermis. They can be congenital or acquired, and most cases are congenital. CSMHs develop from excessive production of smooth muscle at the time of maturation of the mesoderm during fetal life, and acquired smooth muscle hamartomas have been reported most frequently in association with a Becker's nevus[3,8,9].

CSMH has been regarded as excessively rare. But, this once rare lesion is now recognized with increased frequency. Recent reports suggest the estimated prevalence is about 1:2,700 to 1:2,600 live births. This condition does not appear to be so uncommon, but it might be easily overlooked by physicians who are unfamiliar with this lesion[1,8,10].

Lesions are generally single. The most common locations are trunk, especially lumbosacral area, and it also occurs on the upper and lower extremities[1,2,11].

Three or four clinical variants of CSMH have been described by different authors[8,12]. Gerdsen et al.[12] proposed a clinical classification of CSMH into four types. Type 1 is the most common, classical localized type that shows a hairy, hyperpigmented plaque and positive pseudo-Darier's sign. Type 2, a less common subentity of CSMH is characterized by a circumscribed annular patch with multiple follicular papules. There is no hyperpigmentation, and the hair pattern changes are less prominent. Our case belongs to this clinical type. The other types are multiple CSMH and diffuse CSMH, respectively.

More or less than 60% of lesions have been lightly to darkly pigmented and 40% of lesions flesh-colored[2]. The pseudo-Darier's sign, present in CSMH, is a transient induration of the skin that appears when the lesion is rubbed. The induration is the result of the smooth muscle contraction that is due to stimulation of the arrector pili muscles. A positive pseudo-Darier's sign is present in up to 80% of cases, and may be helpful to the differential diagnosis of the disease[1,13].

Histopathologically, numerous thick, well-defined bundles of smooth muscle fibers are scattered throughout the dermis and extend in various directions. Smooth muscle bundles can be readily distinguished from collagen by the trichrome stain. In patients with hypertrichosis, some of the smooth muscle bundles show connections with large hair follicles[2,4,14].

Our present case showed a skin-colored patch with prominent follicular papules. There was no hyperpigmentation or hypertrichosis. The pseudo-Darier's sign was negative. Histopathologic examination revealed typical findings of CSMH. These clinical features belonged to the less common clinical type of the disease[2,8,9].

CSMH may be grossly similar to Becker's nevus. But, CSMH appears at birth, having prominent vellus hairs in those cases with hypertrichosis. In CSMH, the hyperpigmentation, skin induration, and hypertrichosis diminish over time. Becker's nevus, in contrast, appears at childhood or puberty, and hyperpigmentation always precedes hypertrichosis containing thick, terminal hairs, and these signs tend to increase[1,2,7,13]. But, since similar smooth muscle proliferations may be also observed in Becker's nevus, there has been controversy on the relationship between smooth muscle hamartoma and Becker's nevus. Many recent reports suggest that these two entities represent different poles of the same developmental spectrum of hamartomatous change[1,15,16].

It is most important to distinguish CSMH from congenital pigmented nevi prior to surgical treatment. These two entities can be easily differentiated from each other histopathologically. CSMH has not been associated with malignant change, therefore, unnecesary surgical treatment other than for cosmetic reasons is of no therapeutic value and may result in unnecessary permanent deformity[1,2,13,17].

We report herein a case of CSMH showing a distinctive patchy follicular accentuation with no hyperpigmentation or hypertrichosis. This variant is the less common clinical type of the disease, and, to our knowledge, it has not been described in Korean dermatologic literature as yet.

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