Sir,

Congenital smooth muscle hamartoma (CSMH) is an organoid malformation, involving epidermal and dermal structures. The predominant feature is hyperplasia of the dermal smooth muscles.

We present 2 cases of CSMH and describe the condition and all its variants.

CASE REPORTS

Case 1
A 2-year-old girl presented with an asymptomatic congenital skin lesion on her right upper arm. Examination revealed an elevated, slightly hyperpigmented plaque, 6 × 3 cm in size. There was prominent overlying hair and a localized folliculitis that had developed 6 months earlier (Fig. 1). The pseudo Darier sign was positive, with visible piloerection and increased firmness produced by rubbing the lesion.

Histopathology showed a prominent dilated hair follicle with a keratin plug and a perifollicular inflammatory infiltrate with some mucin. Multiple bundles of smooth muscles, running in various directions, were demonstrated in the surrounding dermis.

Four months later, after use of a topical antibiotic preparation, the folliculitis had decreased in intensity. The rest of the lesion remained unchanged.

Case 2
A 48-year-old man was referred with an asymptomatic skin lesion on his left shoulder. On examination there was a circumscribed patch, 20 × 20 cm in size, with firm follicular papules (Fig. 2). According to the patient these asymptomatic lesions had been there since birth. There was no hyperpigmentation and the pseudo Darier sign was negative.

Histology showed numerous bundles of smooth muscles in the mid-dermis, mostly unattached to the hair follicles (Fig. 3).

DISCUSSION

In 1923 Stokes reported a naevus pilaris with hyperplasia of non-striated muscle (1). This condition has since been given various names (2, 3). Internationally, the term congenital smooth muscle hamartoma of the skin (CSMH) is now accepted.

The classical localized variant of CSMH shows a unilateral, hairy, hyperpigmented plaque on the trunk or on a proximal limb (2, 3). Rubbing the lesion leads towards piloerection and a transiently increased firmness (pseudo Darier sign) (3).
Another subentity of CSMH is characterized by a circumscribed annular patch with multiple follicular papules. Unlike classical CSMH, there is no hyperpigmentation and the hair pattern changes are less prominent (4).

There have also been reports of patients with multiple CSMH (5).

Diffuse CSMH has been described as part of the Michelin-tyre-baby syndrome (6). This syndrome is characterized by a combination of anomalies, such as ringed skin creases, cleft palate, epicanthal folds, hypertelorism, malformed ears and developmental delay (6).

On the basis of these criteria, we propose a clinical classification of CSMH of the skin.

1. Type 1: classical localized CSMH
2. Type 2: patchy follicular variant
3. Type 3: multiple CSMH
4. Type 4: diffuse CSMH

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