Congenital Smooth Muscle Hamartoma of the Skin: Clinical Classification

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).

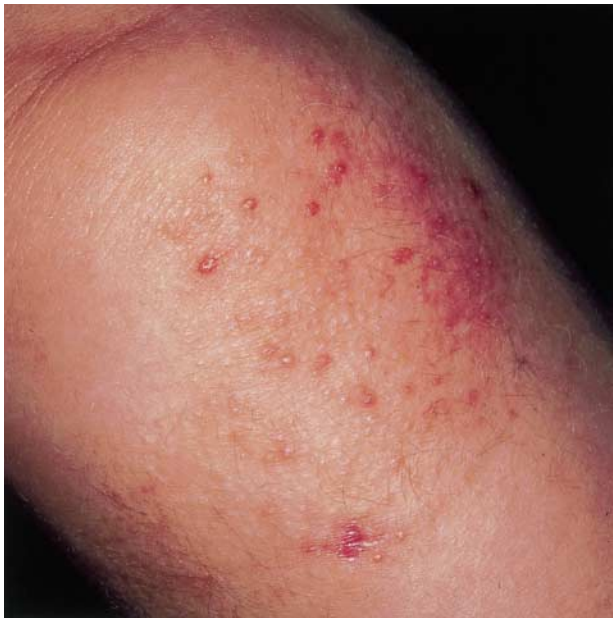

Fig. 1. Case 1: classical localized CSMH on the right upper arm of a 2-year-old girl.

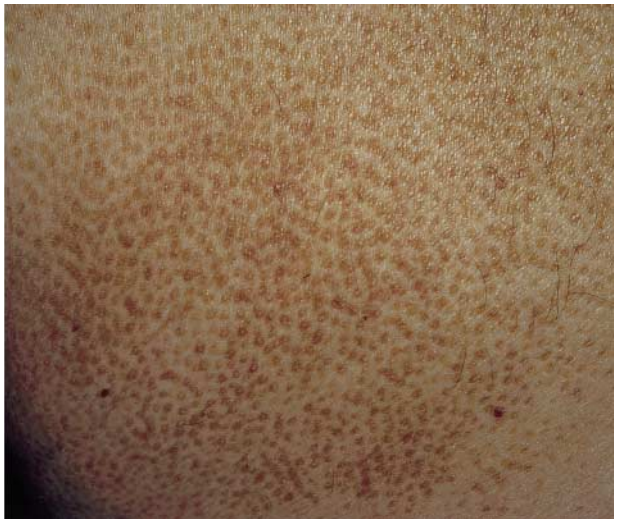

Fig. 2. Case 2: patchy follicular variant of CSMH on the left shoulder of a 48-year-old man.

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).

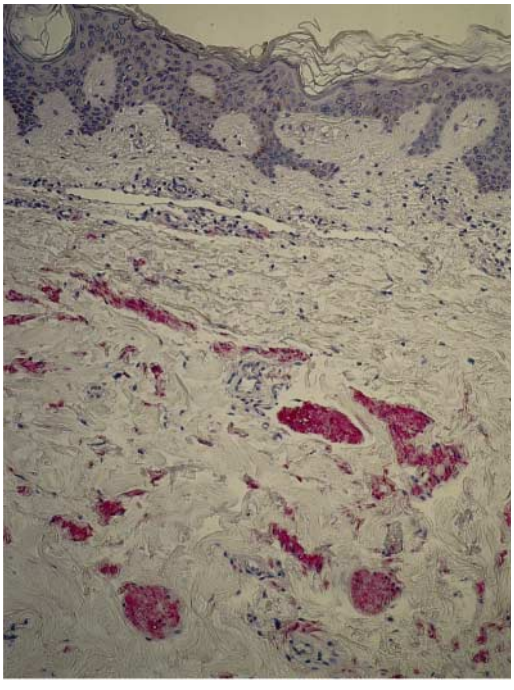

Fig. 3. Case 2: immunohistochemistry (Desmin, clone D33); proliferation of smooth muscle bundles in the dermis.

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, hyperteleorism, 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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Risk Factors for Skin Cancer in a Group of Renal Transplant Recipients

Sir,

Studies in northern Europe and Australia have detected a high incidence of skin cancer in renal transplant recipients (RTRs). Hartevelt et al. (1) found a cumulative incidence of skin cancer, ranging from 10% to 40%, at 10 and 20 years after transplantation. Bouwes Bavinck et al. (2), in a cohort of Australian patients, reported a higher incidence (from 45% to 70% at 10 and 20 years after transplantation), related to the more intense sun exposure at those latitudes. Only few reports have been published about RTRs from southern Europe and Italy. From 1990 to 1997 we enrolled a consecutive series of RTRs in a dermatological screening program, followed up by the Second Division of Surgery and Kidney Transplantation Centre of the Ospedale Civile Maggiore, Verona, Italy. All patients gave their informed consent before medical examination.

We examined 423 RTR subjects (290 males and 133 females), treated with 3 different immunosuppressive regimens: prednisolone + azathioprine (PA) (71 patients), prednisolone + azathioprine + cyclosporin (PAC) (191 patients), prednisolone + cyclosporin (PC) (161 patients). An accurate dermatological anamnesis was collected. Clinical records were available for all the patients. For each patient we recorded the following data: age, sex, stature, weight, date of the transplantation and of the visit, type and maintenance posology of the immunosuppressive drugs. The whole skin surface and mucous membranes were examined by a dermatologist. Any lesion suspected to be a skin cancer was excised and its histology examined. For estimates of potential prognostic factors, only variables available at the time of the transplant were considered: we took as an end point the diagnosis of cancer. Cox's proportional-hazards regression model (3), performing backward stepping of variables with pre-assigned p values equal to 0.05 that controlled the stepping removal, and Kaplan and Meir survival curves were carried out.

The mean age of the RTRs at the visit was 46.2 years (SD 11.2, age range 19–68 years) and mean age at transplantation was 38.9 years (SD 11.7 age range 12–65 years), with no difference between males and females. Mean follow-up time was 7.6 years (SD 5.38, range 0–26 years): the PA group had a longer follow-up time (14.0 years) (SD 6.0) than the PAC group (6.6 years) (SD 3.9) and the PC group (5.9 years) (SD 4.7).

A total of 43 patients were excluded: 30 died of various accidents unrelated to the graft function and 13 rejected the graft (Table I).

A skin cancer was detected in 21/423 patients (5%) (BCC 10; Bowen 2; Keratoacanthomas 3; SCC 6). Cumulative incidence of the first cancer at 5, 10, 15 and 20 years of follow-up was 0.8%, 5.2%, 11.2% and 15.3%, respectively. The incidence of cancer was not significantly different in the 3

Table I. *Patients excluded from the study*

Immunosuppressive therapy	Death	Rejection	Total
PA	9	5	14
PAC	7	4	11
PC	14	4	18
Total	30	13	43

PA = prednisolone + azathioprine.
PAC = prednisolone + azathioprine + cyclosporin.
PC = prednisolone + cyclosporin.