Patchy Follicular Spotted Macules on the Lower Legs

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CASE REPORT

A 73-year-old female was found to have numerous follicular spotted macules on right lower leg, and a few macules on bilateral ankles for one year (Fig 1.). These macules first began on right ankle and extended slowly up to the right knee. They were asymptomatic, red to brown in color. The patient denied any known systemic disease and drug allergic history. Under the impression of vasculitis, a skin biopsy on right knee was performed. The histopathologic examination revealed dermal proliferation of smooth muscles bundles with focal superficial perivascular infiltrate. Mild basal hyperpigmentation and elongation of rete ridges were also noted (Fig 2.). The skin lesions did not improve after treatment with topical corticosteroids and hydroquinone for 3 months. A second skin biopsy on right ankle was performed. The histopathologic findings were similar.

Fig. 1
A 73-year-old female with patchy follicular, spotted macules on her right lower leg.

Fig. 2
H & E, 40X
DIAGNOSIS: Acquired Smooth Muscle Hamartoma

DISCUSSION

Smooth muscle hamartoma (SMH) is a benign proliferation of smooth muscle bundles in the dermis. Normal distribution of cutaneous smooth muscle is presented in 3 locations: arrector pili muscles, blood vessel walls, and dartos tunica/areola. Proliferation of atopic smooth muscle is regarded as hamartoma. SMH was first reported by Strokes in 1923. It can be congenital or acquired.

Congenital SMH developed during fetal life and clinically appeared as skin-colored or variable pigmented patches or plaques with or without hairs, usually located on the trunk, extremities, especially the lumbosacral area. The prevalence is about 1:1,000 to 1:2,700 of live birth with a slight male predominance.

Acquired SMH was reported most frequently in association with Becker’s nevus, which is an organoid hamartoma that contains atopic hairs, muscles and nerves. Becker’s nevus appeared in the first or second decade, often located on the shoulder, chest. The appearance of hyperpigmentation usually preceded hypertrichosis. Although some authors considered SMH as a distinct clinicopathologic entity, it is more common to regard that SMH and Becker’s nevus are two polar entities at either end of a continuous spectrum of dermal smooth muscle proliferative disorders.

SMH with patchy follicular spotted maculopapules was a rare clinical variant and all the previous reported 6 cases were congenital. The follicular macules were skin to red colored in 3 cases and pigmented in 3 cases. Smooth muscle bundles in these specimens were not connected with hair follicles. To our knowledge, our case is the first case presenting as acquired follicular macular pigmentation, as the patient is an old-aged woman with skin lesion of one year. Histologically, both sites of skin biopsies revealed dermal proliferation of smooth muscles bundles with focal superficial perivascular infiltrate. Mild basal hyperpigmentation and elongation of rete ridges were also noted.

The clinical differential diagnosis of our case includes congenital pigmented hairy nevus, cafe-au-lait spot, connective tissue nevus, Becker’s nevus, macular or lichen amyloidosis, ichthyosis, ataseotatic dermatitis or follicular eczema with postinflammatory hyperpigmentation. Pseudo-Darier’s sign in SMH can be elicited, but not always. Masson’s trichrome stain was used to confirm the smooth muscle of origin. Leimyoma is histologically confused with SMH that the latter has a well-defined, scattered of smooth muscle proliferation, where the proliferative smooth muscles in leimyoma are ill-defined, and tightly interlaced. Leimyoma usually appears as painful nodules.

Extensive lesions of SMH associated with systemic disease or underlying developmental abnormalities was reported in Michelin-tire syndrome. No malignant transformation of the skin lesion was noted. No treatment is necessary except for the cosmetic concern.

REFERENCES