

## Hairy plaque on the thigh

A 23-year-old woman presented to our institution with a solitary, slightly elevated, hairy plaque on the extensor surface of her left thigh. It had been present since birth and had gradually increased in size. She stated that after rubbing or scratching, the lesion would raise and harden temporarily; but otherwise, it was asymptomatic. The patient's medical and family histories were unremarkable.

Physical examination revealed an ill-defined 10×5-cm skin-colored plaque with prominent overlying hairs and confluent, rubbery perifollicular papules [Figure 1]. A 4-mm punch biopsy of the lesion was obtained and stained with hematoxylin-eosin [Figure 2], and Masson's trichrome [Figure 3].

### WHAT IS YOUR DIAGNOSIS?



Figure 1: Skin-colored plaque with overlying hairs and confluent, rubbery perifollicular papules

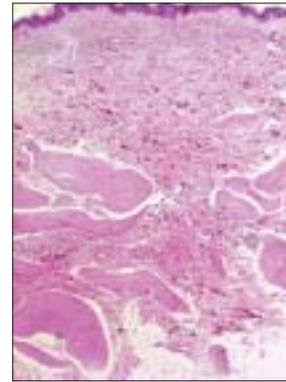


Figure 2: Scattered smooth muscle bundles in the dermis (H and E, ×40)



Figure 3: Masson trichrome stain (×40)



Figure 4: The antibody for smooth muscle actin confirmed the presence of smooth muscle fibers (immunoperoxidase, ×40)

### **Diagnosis: Congenital smooth muscle hamartoma**

Histopathological examination revealed a normal epidermis and haphazardly distributed smooth muscle bundles in the reticular dermis. A thin retraction space around the bundles separating them from the adjacent dermal collagen also was observed [Figure 2]. The smooth muscle bundles were clearly seen with Masson's trichrome stain [Figure 3]; these also stained positively with smooth muscle actin [Figure 4].

## **DISCUSSION**

Congenital smooth muscle hamartoma (CSMH) is a skin-colored or lightly pigmented patch or plaque with hypertrichosis that originates from the smooth muscle fibers of the erector pili. It is often present at birth, with an estimated incidence of 1 in 2600 live births, but it also may be acquired.<sup>[1]</sup> It presents as a solitary lesion on the trunk or proximal extremities, with involvement of the lumbosacral area in two thirds of the patients.<sup>[2]</sup> Rubbing the lesional skin causes transient elevation or induration called the pseudo-Darier sign, which may be seen in 80% of the patients.<sup>[3]</sup>

CSMH is characterized histologically by proliferation of smooth muscle bundles of the erector pili muscles in the reticular dermis. The pathogenesis is unknown. It may be due to the excessive proliferation of smooth muscle fibers at the time of maturation of the mesoderm during fetal life.<sup>[4]</sup> Abnormal proliferation of CD34-positive dermal dendritic cells is supposed to stimulate aberrant development of smooth muscle proliferation by releasing growth factors.<sup>[5]</sup>

The clinical differential diagnosis of CSMH includes congenital melanocytic nevus, Becker's melanosis, solitary mastocytoma, leiomyoma, connective tissue nevus, nevoid hypertrichosis, and acquired localized hypertrichosis.<sup>[3]</sup> Differentiating CSMH from congenital melanocytic nevus is critical, as the latter, especially the giant congenital melanocytic nevus, may have a malignant potential. In congenital melanocytic nevus, a pseudo-Darier's sign is not observed; and histopathologically, there is no smooth muscle proliferation in the dermis. In Becker's melanosis, hyperpigmentation and hypertrichosis tend to increase with

time; while in CSMH, these are not the predominant features, and they tend to decrease with age. Histopathologically, there is no smooth muscle proliferation in the dermis, and epidermal features such as increased pigment in the basal layer, acanthosis, elongation, and clubbing of the rete ridges are observed more frequently in Becker's melanosis than in CSMH. Solitary mastocytoma and connective tissue nevus can be easily distinguished from CSMH by the presence of mast cell infiltration and increased dermal collagen respectively.

Congenital smooth muscle hamartoma has a good prognosis with no reports of malignant transformation. Although no treatment is necessary, if the CSMH is cosmetically bothersome, surgical excision is the best option. Shaving, laser therapy, and chemical epilation are other treatment options.

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