

Sir,

A smooth muscle hamartoma is a benign proliferation of smooth muscle bundles within the dermis.<sup>[1]</sup> Most lesions are congenital and acquired lesions have rarely been reported.

A 32-year-old male presented with increased hair growth on the left side of upper back of 6 months duration. There was no history of itching or pain in the lesion. Cutaneous examination revealed a large patch of hypertrichosis with terminal hair on the left side of upper back in the scapular area [Figure 1a]. On close observation, the skin in the hypertrichotic area was normal in color. However, on shaving, slightly hyperpigmented small follicular papules were noted [Figure 1b].

A punch biopsy taken from the hairy area showed mild epidermal hyperplasia with increased basal layer pigmentation. Dermis showed multiple bundles of smooth muscle cells arranged haphazardly in the reticular dermis [Figure 2a]. On Masson trichrome stain, these bundles stained red in color [Figure 2b].

Smooth muscle hamartoma arises from smooth muscle cells that are located in arrector pili muscle, dartos muscle, vascular smooth muscle, muscularis mammillae, and the areolae.<sup>[2]</sup> Acquired smooth muscle hamartoma is a rare entity. Most of the cases of acquired smooth muscle hamartoma were shown

to have originated from arrector pili and dartos muscles.<sup>[1]</sup> Acquired smooth muscle hamartoma is usually located on the skin of the forearm, chest, neck, scrotum, penis, labia majora, or shoulder.

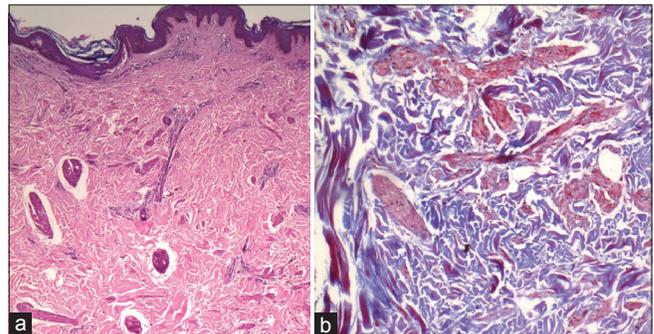
Congenital smooth muscle hamartoma usually manifests at birth, but rarely, can also appear later in life. It is usually located on the trunk and extremities and appears as variably-sized papules, patches, or plaques.<sup>[3]</sup> Pseudo-Darier's sign is considered as the characteristic diagnostic clue for congenital smooth muscle hamartoma.<sup>[1]</sup> Pseudo-Darier's sign is a transient piloerection and elevation or induration of lesional skin, while Darier's sign refers to the urtication and erythematous halo which is seen in response to stroking or rubbing of the lesional skin in cutaneous mastocytosis.<sup>[4]</sup>

In contrast to congenital smooth muscle hamartoma, acquired smooth muscle hamartoma appears later in life as a slightly hyperpigmented or hypertrichotic patch or plaque which may be associated with itching, pain, or numbness. Pseudo-Darier's sign is usually negative in these lesions.<sup>[5]</sup>

The histopathologic hallmark of smooth muscle hamartoma is an increase of large, randomly oriented bundles of smooth muscle cells with central, cigar-shaped nuclei. These smooth muscle bundles are arranged haphazardly in the dermis.



**Figure 1: (a) A patch of hypertrichosis over the scapular area (b) Follicular papules which were revealed after shaving**



**Figure 2: (a) Haphazardly arranged multiple bundles of smooth muscle cells in the dermis (H and E, x100) (b) Smooth muscle bundles stained red (Masson's trichrome, x200)**

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Clinically and histologically, Becker's nevus often resembles smooth muscle hamartoma. Becker's nevus (also known as Becker's pigmented hairy nevus) presents as a well defined, uniformly hyperpigmented plaque with hypertrichosis and thickened affected skin that usually manifests at puberty. Histopathology shows epidermal hyperplasia with flattening at the bottom of rete ridges and increased basal layer pigmentation. Occasionally, Becker's nevus can also show a proliferation of dermal smooth muscles.<sup>[6]</sup> These clinical and histopathological features help to distinguish Becker's nevus from acquired smooth muscle hamartoma. Because of the overlapping histopathological features, some authors have speculated that Becker's nevus and smooth muscle hamartoma are two polar entities at either end of a continuous spectrum.<sup>[3]</sup>

As acquired smooth muscle hamartoma is a benign condition, no treatment is usually required. However, for cosmetic reasons, laser hair removal and surgical excision may be attempted.

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