

Cutaneous smooth muscle tumors

by Hayder A. Asfoor, MD

Smooth muscle hamartoma

- Congenital (most common) or acquired
- Clinically:
 - Firm, hyperpigmented or skin-colored plaque ± hypertrichosis or follicular papules
 - Diffuse cutaneous smooth muscle hamartomas may result in the Michelin-Tire Baby phenotype.
- May be associated and overlap with Becker's nevus clinically and on histopathology. However, smooth muscle hamartomas classically differ from Becker's nevus by earlier onset (at birth vs second decade) and location (trunk and extremities vs shoulder or chest).
- May be pseudo-Darier's sign positive (stroking the lesions cause induration and erythema due to SM contraction).
- Histology→ Haphazardly arranged SM bundles in the dermis with epidermal acanthosis and basal layer hyperpigmentation.
- IHC→ positive for SMA, desmin, and smoothelin
- Tx: May perform excision for cosmetic reasons (no malignancy potential)

Leiomyoma

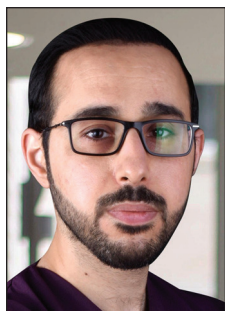
- Benign mesenchymal neoplasms with SM differentiation
- Three Types:

1. Pilar Leiomyoma (arrector pili origin)

- Two Types:
 - Multiple: younger age of onset affecting the trunk and limbs equally. a/w Reed's syndrome (+ uterine leiomyoma and RCC)
 - Solitary: typically in adults, with affinity for the limbs.
- Clinically→ firm, reddish-brown or skin colored nodules, papulonodules, or plaques; typically painful, especially with cold exposure
- May be pseudo-Darier's sign positive
- Histology→ circumscribed proliferations of interlacing bundles of SM fibers within the reticular dermis
- IHC→ positive for SMA and desmin. Calponin and h-caldesmon are also supportive
- Tx:
 - Solitary: complete surgical excision
 - Multiple: nifedipine, nitroglycerin, phenoxybenzamine, gabapentin and hyoscine hydrobromide for pain, CO2 laser

2. Genital Leiomyoma (Dartocic, vulvar or mammary sm origin)

- Clinically→ similar to pilar type but usually larger; typically solitary and painless
- Involving vulva, scrotum, penis, nipple, or areola
- Histology→ more circumscribed appearance than pilar leiomyomas and may have mitoses
- IHC and Rx: same as pilar leiomyoma



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Leiomyoma (cont.)

3. Angioleiomyoma (perivascular SM origin)

- Pericytic tumor (Myoid differentiation)
- Clinically→ solitary nodule in the lower extremities; often painful
- Histology→ well circumscribed proliferation of fascicles of spindled SM cells admixed with blood vessels within SC. Three variants: solid, venous, and cavernous.
- IHC→ positive for MSA, SMA, and calponin. Variable for desmin and h-caldesmon.
- Tx: Simple surgical excision

Leiomyosarcoma

- Rare: ~4% of cutaneous soft tissue sarcomas.
- Two types:
 - Dermal leiomyosarcoma: arises from arrector pili or genital sm with indolent clinical behavior
 - Subcutaneous leiomyosarcoma: arises from vascular sm with aggressive behavior and risk of metastasis in up to 40%
- Clinically→ solitary red brown and painful nodule, involving extremities, trunk, and head and neck region of elderly
- Histology→ diffusely infiltrative or well-demarcated nodular pattern with SM differentiation + variable degrees of atypia
- IHC→ positive for MSA, SMA. Variable for desmin (though classically +), h-caldesmon, keratin and S100.
- Tx: WLE

Abbreviations:

- IHC: immunohistochemistry
- SMA: smooth muscle actin
- MSA: muscle specific actin
- a/w: associated with
- RCC: renal cell carcinoma
- SM: smooth muscle
- SC: subcutis
- Tx: treatment
- WLE: wide local excision

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