Congenital smooth muscle hamartoma

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Congenital smooth muscle hamartoma. ORPHA:263435

Congenital smooth muscle hamartoma (CSMH) is a rare cutaneous hamartomatous lesion most often located on the lumbosacral area or proximal limbs (but rarely on atypical areas such as scalp, eyelid or foot) and characterized by a disorganized proliferation of smooth muscle fibres of arrector pili presenting usually as a localized skin-colored or hyperpigmented plaque (up to 10 cm in diameter) with prominent vellus hairs (most common classic form) or less commonly by multiple skin-colored papules that can coalesce to form irregularly shaped plaques. With time, hyperpigmentation and vellus hairs usually diminish and neither malignant transformation nor associated systemic involvement has been reported.