Hyperpigmented patch on the trunk of a neonate

A one-day-old, full-term, normally delivered, female neonate was being routinely examined. A brownish irregular patch of 2 cm × 1.5 cm, with fine long hairs was observed on her right loin [Figure 1]. On gentle stroking, the lesion became more prominent along with the appearance of goose bumps. A punch biopsy was taken from the edge of the lesion. Histopathological preparation on H/E stain showed mild hyperkeratosis of the epidermis and basal cell hyperpigmentation. In the reticular dermis, well-defined smooth muscle fiber bundles were seen, interspersed among hair follicles [Figure 2]. Masson Trichrome stain [Figure 3] confirmed the presence of smooth muscle fibers (stained red).

WHAT IS THE DIAGNOSIS?
Answer to Quiz

Diagnosis: Congenital smooth muscle hamartoma (CSMH) or congenital arrector pili hamartoma.

DISCUSSION

CSMH was first described by Stokes in 1923.[1] The lesions may be solitary or multiple, the former being commoner (1 in 3000 births).[2] Classically, it is present at birth or seen during the first week of life, located most commonly over the lumbo-sacral region.[2] Other sites of occurrence are buttocks, proximal extremities, and rarely the scalp.[3] Multiple lesions are rare and may occur in a linear pattern.[4,5] Diffuse involvement may occur with the Michelin type baby syndrome.[6]

The commonest morphological pattern is a skin-colored or slightly hyperpigmented patch or plaque associated with long vellus hairs or small follicular papules all over the lesion.[2] On gentle stroking, the lesion shows vermiculation and pilo-erection (Pseudo-Darier’s sign)[2,7] or occasionally fasciculation.[2] Lesions occurring as a linear atrophic plaque[4] and a patch with perifollicular papules[5] have been reported.

CSMH is related to the arrectores pilorum muscle,[8] which in fetal life originates from a diffuse metachromatic zone of mesoderm situated near the hair germ. Over-proliferation of this region is likely to give rise to the lesion. Clinically, CSMH has to be differentiated from café-au-lait macule, solitary mastocytoma, congenital melanocytic nevus and connective tissue nevus.[2] Histopathologically, pilar leiomyoma is an important differential diagnosis.[8] A well-defined bundle of long, straight smooth muscle cells in the deeper dermis, which are separated from the dermal collagen by a clear space, is characteristic of CSMH. Irregular smooth muscle bundles, intermingled with collagen, which may be attached to the hair follicle, are found in pilar leiomyoma.[5,8]

An acquired form of smooth muscle hamartoma without hyperpigmentation or excessive hair has been reported.[9] Other acquired lesions occur in association with Becker’s nevus.[4] There are clinical and histopathological overlaps between Becker’s nevus and CSMH.[7,8] Some authors include both the entities in a spectrum of hamartomatous disorders,[7] involving the epidermis (hyperpigmentation), hair follicles (hypertrichosis) and the dermis (smooth muscle). Children with the diffuse form of the disease may have other congenital malformations, growth and mental retardation.[7] No treatment is necessary for solitary lesions, which may persist unchanged or there may be slight reduction of the hypertrichosis.

Aparna Palit, Arun C. Inamadar, S. B. Athanikar, V. V. Sampagavi, N. S. Deshmukh, B. R. Yelikar* Department of Dermatology, Venereology and Leprosy, and *Pathology, BLDEA’s SBMP Medical College, Hospital and Research Centre, Bijapur, India

Address for correspondence: Dr. Arun C. Inamadar, Department of Dermatology, Venereology and Leprosy, BLDEA’s SBMP Medical College, Hospital and Research Centre, Bijapur - 586103, Karnataka, India.
E-mail: aruninamadar@rediffmail.com

REFERENCES