Case report

Congenital smooth muscle hamartoma on the scalp

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Case Report

An 8-day-old male infant was referred to our neonatal unit for prematurity. It was observed that his scalp color was darker than the rest of his skin color when the scalp hair was partially shaved for intravenous access. Dermatologic examination revealed a slightly elevated, hyperpigmented plaque confined to the whole scalp. The margin of the lesion did not extend to the face and neck. His hair was abundant and dark (Fig. 1). There were no areas of alopecia and no overlying folds of skin. There were no dysmorphic features, such as Michelin tire baby. Pseudo-Darier’s sign was negative.

A punch biopsy specimen was obtained from the lesion with a diagnosis of congenital melanocytic nevus or congenital blue nevus. The result of histopathologic examination showed increased smooth muscle bundles in the reticular dermis running in various directions under a normal epidermis (Fig. 2a); immunohistochemical stain revealed desmin-positive (Fig. 2b) and S100-negative proliferation. Based on these clinical and histopathologic findings, a diagnosis of congenital smooth muscle hamartoma was made. No changes in this lesion were observed during a 9-month follow-up period.

Discussion

Congenital smooth muscle hamartoma (CSMH) is a rare cutaneous dysembryoplasia characterized by a disorganized proliferation of normal muscle fibers of arrector pili. The disease usually presents as a localized, skin-colored or mildly hyperpigmented, irregularly shaped patch or plaque with prominent vellus hairs located on the lumbosacral area.

Figure 1 Congenital smooth muscle hamartoma on the scalp of an 8-day-old premature newborn
Malignant transformation has not been reported and pigmentation diminishes with time.\(^1\)\(^2\) The prevalence of CSMH has been estimated to be 1 in 2,600 live births, with a slight male predominance;\(^3\) however, recent reports have suggested that CSMH is probably more prevalent than indicated by the small number of cases reported in the literature.\(^4\)

Although excessive hairiness is the most frequent sign of CSMH, lesions in the head and neck region have rarely been reported in the literature. Only two reports have described CSMH on the head, one of which was located on the frontal scalp and the other on the face.\(^4\)\(^5\) In our case, the lesion appeared over the whole scalp and, interestingly, the margin of the lesion did not cross the line of the scalp hair. We noticed the lesion during shaving for intravenous access, otherwise it might have remained unrecognized.

The diagnosis of CSMH is based on histopathologic examination. Histology is characterized by the presence of numerous smooth muscle fibers disseminated in the dermis and diversely oriented, sometimes in contact with hair follicles which retain their normal morphology. The clinical differential diagnosis of CSMH includes congenital melanocytic nevus, Becker’s nevus, solitary mastocytoma, piloleiomyoma, café-au-lait spots, nevus pilosus, and occult spinal dysraphism.\(^2\)\(^5\)

Distinguishing CSMH from congenital melanocytic nevus may be difficult on clinical grounds. Congenital melanocytic nevi are differentiated from CSMH by histopathologic examination. The differentiation of these two clinical pictures is very important for the follow-up of patients, as congenital melanocytic nevi carry the risk of malignant transformation. Becker’s nevus is another important entity in the differential diagnosis of CSMH. It resembles CSMH both clinically and histopathologically. In contrast with CSMH, Becker’s nevus is usually an acquired lesion, although congenital cases have been reported very rarely.\(^2\)\(^6\)

We also considered giant congenital blue nevus in the clinical differential diagnosis of our patient. Giant congenital blue nevus of the scalp presents with local invasion of muscle, bone, and meninges in most cases, with a risk of malignant transformation. We differentiated this clinical entity from CSMH by histopathologic examination, which revealed no spindle-shaped or dendritic cells with melanin granules in the dermis, typical of blue nevus. Immunohistochemical staining of the biopsy specimen from our patient was also S100 negative.

The recognition and differentiation of CSMH from congenital melanocytic nevus is particularly fundamental to avoid unnecessary excision of this benign lesion. It is important to emphasize that giant congenital blue nevus of the scalp should also be included in the differential diagnosis of CSMH when the lesion is located on the scalp. CSMH lesions located on the scalp may be ignored and thus their frequency may be underestimated.

References


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**Figure 2** (a) Increased smooth muscle bundles in the reticular dermis running in various directions under the epidermis (hematoxylin and eosin; magnification, \(\times 100\)).

(b) Desmin positivity (immunohistochemistry, \(\times 200\))
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