Hair Collar Sign Associated with Scalp Aplasia Cutis Congenita

E Roche-Gamón, I Febrer-Bosch, and V Alegre de Miquel
Servicio de Dermatología, Hospital General Universitario, Valencia, Spain

To the Editor:

The term “hair collar sign” was first introduced into the dermatological lexicon by Commens et al1 in 1989. The sign consists of a ring of long, dark, thick, and rough hair surrounding a congenital nodule of cystic, blister-like, or atrophic appearance located on the scalp. Histology reveals numerous horizontally oriented hypertrophic hair follicles emerging from the edge of the lesion. Drolet et al2 were the first to highlight the importance of the hair collar sign as a marker of spinal dysraphism. They proposed that its formation could be caused by cerebral herniation that would produce, early in embryonic development, abnormal shearing forces during formation of hair follicles, causing them to point outward from the defect. The proximity of the neuroectoderm, which expresses neural cell adhesion molecules, could also alter normal dermal-epidermal interactions, and as a consequence induce the development of large abnormal follicles.

We present the case of a newborn child from healthy parents. The mother was monitored during her pregnancy, had no adverse events, and had not taken medications. The delivery was vaginal, without the use of instruments. At birth, the child presented a rounded cutaneous defect with a diameter of 1 cm in the left parietal area, near the vertex. The defect was composed of an erythematous, slightly protruding lesion with an edematous appearance and covered by a fine, atrophic, translucent membrane. It was surrounded by abundant thick, dark, rough hairs that were horizontally arranged and oriented toward the periphery of the lesion (Figure 1).

There were no palpable underlying bone abnormalities. The infant also presented a symmetric defect on both hands involving duplication of the thumb, which also presented syndactyly (Figure 2). Ultrasound of the brain and through the fontanelle ruled out abnormalities of the bone or nervous tissue. We finally established

References


Figure 1. Hypertrophic hair collar surrounding the cutaneous defect.
the diagnosis of membranous aplasia cutis congenita (ACC) of the scalp with hair collar sign, associated with polydactyly and syndactyly. This diagnosis could correspond to ACC group 1 of the scalp with no associated abnormalities or associated with isolated anomalies.\textsuperscript{3}

Two aspects should be mentioned in relation to this case. First of all, a comprehensive assessment should be performed on any congenital lesion of the midline before surgical excision or biopsy. Computed tomography is the most accurate method for assessing cranial defects. Any bone defect observed with this method should be examined by magnetic resonance imaging to determine if there is transcranial extension of soft tissue. If there is no cranial defect and the lesion is consistent with membranous aplasia cutis, then biopsy is not necessary. Secondly, the association of various cutaneous markers on the scalp, such as the hair collar sign and vascular malformations, increases the possibility of associated cervical dysraphism.\textsuperscript{4,5}

References