Hair Collar Sign Associated with Scalp Aplasia Cutis Congenita

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To the Editor:

The term “hair collar sign” was first introduced into the dermatological lexicon by Commens et al 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 al 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...
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.  

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.  

To the Editor:

In dermatological pathology, a panoramic view of the histological specimen being examined is important. A panoramic histological view allows assessment of the localization of the skin condition, as well as identification of certain inflammatory or tumor patterns and/or architecture that may aid the diagnosis.  

A panoramic view of a histological specimen will require a photographic camera fitted to a microscope that, in addition to the usual lenses (4×, 10×, 20×, 40×, and 100×), has a 1.25× or 2× zoom lens or magnifying lens. A lens of this type is often expensive and not always available. As a result, adequate images are often missing from most dermatopathological publications and reports.  

A course on diagnosis in dermatological pathology was recently taught by Dr Requena and Dr Sánchez-Yus in Madrid, Spain, in which emphasis was placed on the importance of panoramic views of dermatological lesions for diagnosis. Some of the panoramic histopathological images shown in the course had been taken with a single-lens reflex camera directly focused on the slide. Our department is equipped with a DermLite FOTO37 dermatoscope coupled to a Nikon Coolpix 4500 camera, leading us to consider using that equipment with a histological specimen to obtain a good panoramic magnified view.  

A dermatoscope is an optical system involving polarized light. The magnification ranges from 10× to 400×, or even more. The dermatoscope was designed to assist in the specific diagnosis of pigmented skin lesions, but has also been used in the diagnosis of other lesions, such as vascular, inflammatory, or parasitic lesions. In rheumatology, the dermatoscope is used by some specialists as a capillaroscope. In dermatology, the dermatoscope is used by some specialists as a capillaroscope.