

Hair Collar Sign with Naevus Flammeus: A Possible Sign of Aplasia Cutis Congenita

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Case Presentation

A 2-month-old female infant presented with a reddish patch and bald area on her scalp, which her parents had noticed one week earlier (Figure 1A). Dermoscopy revealed an atrophic patch lacking central follicular openings, surrounded by radiating coarser hairs and featuring thin linear, rounded, and globular vessels on a pinkish background (Figure 1B). She was diagnosed clinically with aplasia cutis congenita (ACC) with a hair collar sign (HCS) and naevus flammeus, supported by dermoscopic findings. Magnetic resonance imaging (MRI) demonstrated that the skull was closed and intact, with normal intracranial structures (Figure 1C). She was advised to undergo close follow-up without intervention.

bald scalp lesion, termed HCS, is considered a specific clinical sign of ACC. Prior studies suggest nearly half of HCS cases correlate with naevus flammeus (port-wine stains); this association may arise from aberrant migration of neural crest cells affecting fetal skin development, particularly in capillary network formation [1]. In ACC cases, because the presence of HCS and vascular stains are considered as a marker for neuroectodermal defects (e.g., heterotopic brain tissue or cephalocele), an MRI is essential for evaluation [2].

In conclusion, HCS is frequently associated with naevus flammeus and may indicate aplasia cutis congenita. Patients demonstrating this cutaneous marker should undergo thorough physical examination coupled with MRI evaluation of the craniospinal axis.

Teaching Point

Aplasia cutis congenita (ACC) is a rare congenital skin defect that can occur anywhere on the body, most commonly on the scalp. A ring of coarser, darker hair surrounding a

Ethical Statement: Written informed consent was obtained from the patient's parents for publication of this case letter and any accompanying anonymized data/images. The parents were informed that all personal identifiers would be removed to protect privacy.

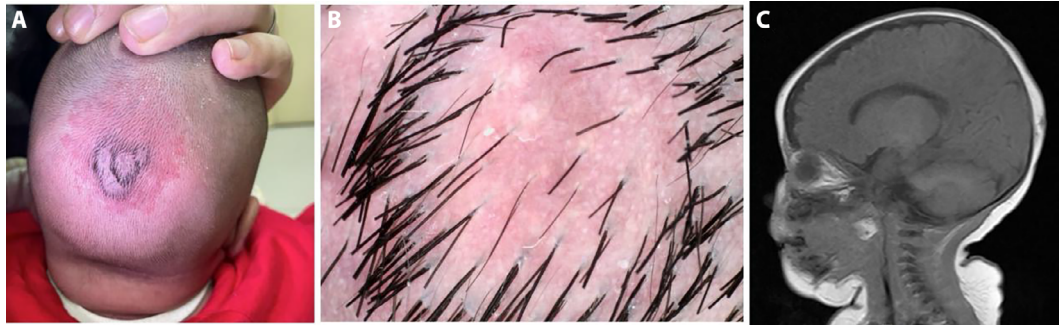


Figure 1. (A) A reddish patch with a hair collar sign is present on the occipital region of an infant; (B) Dermoscopy revealed an atrophic patch surrounded by radiating hair follicles, with thin linear, rounded, and globular vessels on a pinkish background (polarized 30x); (C) MRI scan showed the skull was intact, with normal intracranial structures.

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