IMAGES IN PAEDIATRICS

Aplasia cutis with 'hair collar sign'

A 2-month-old boy was referred to our paediatric dermatological department with two adjacent oval patches of 2×3 cm in diameter, covered by a thin, atrophic, translucent membrane. The larger lesion was surrounded by tufts of darker terminal hair, that were horizontally arranged and oriented towards the periphery of the lesion, starting from the vertex (figure 1). There were no palpable underlying bone abnormalities. The infant was otherwise healthy. A MRI of the brain was normal. The clinical diagnosis was membranous aplasia cutis congenita (ACC) of the scalp with the 'hair collar sign'.

ACC is a rare congenital malformation characterised by well-circumscribed absence of skin that involves the epidermis, dermis and subcutaneous fat.[1 2] The entire skin can be involved, even if the scalp is the most typical site. Usually the skin around the alopecic lesion is not erythematous. The 'hair collar sign' was first described by Commens *et al* in 1989.[3] The sign consists of a ring of long, dark, thick and rough hair surrounding the atrophic alopecic patches on the scalp. This condition may be associated with encephaloceles, meningoceles and heterotopic brain tissue.[4] Recently, dermal melanocytosis and naevus flammeus have also been described in ACC.[2 5] Infants with ACC should undergo a MRI to exclude a transcranial extension of soft tissue, such as spinal dysraphism or a neuroectodermal defect.

In our patient the lesions were unchanged at 6 months follow-up. Small areas of ACC usually heal well with alopecic scars. Larger defects may require surgical intervention, such as skin grafts, local flaps or free flaps, to reduce the risk of infection and bleeding.

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Competing interests None.

Figure 1 Two oval translucent non-inflammatory and well demarcated atrophic patches on the vertex of the scalp (2×3 cm in diameter). Note the growth of long dark coarse hair encircling the larger lesion: the 'hair collar sign'. The rest of the scalp is normal.

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