INTRODUCTION

Membranous, bullous, or cystic aplasia cutis congenita is a clinical subtype of aplasia cutis, covered with a membranous or glistening surface. Trichoscopy may help in the diagnosis.

CASE REPORT

A Caucasian male newborn presented at birth with two flat lesions on the left parietal scalp, surrounded by a rim of terminal hairs [Figure 1]. He was otherwise healthy and was born after an unassisted vaginal delivery and an uncomplicated term pregnancy. At the age of 2 months, physical examination revealed two translucent papules, 4–6 mm [Figure 1]. On dermoscopy (polarized light), the largest showed a reddish background, thin, linear vessels and, remarkably few hair bulbs could be seen because of the translucency of the lesion. No skull bone and brain defects were found. The diagnosis of membranous aplasia cutis congenita was established.

DISCUSSION

Aplasia cutis congenita is a congenital defect of the skin characterized by localized absence of the epidermis, dermis and, at times, underlying structures such as bone or dura.[1] It generally occurs on the scalp. Most cases appear to be sporadic although some potential associations have been proposed: drugs, underline embryologic malformations, infarction caused by rapid growth, between others.[2] The clinical picture varies from fissure-like ulcers with a granulating base, erosions, atrophic macules, or scars. Among them, a cystic variant covered with a membranous or glistening surface is called membranous aplasia cutis congenita (MACC).[3]

In a recent review, all the cases were located on the skull, most of them on the vertex or parietal scalp, ranging...
from 1 to 7 lesions, and frequently associated with bone defects (6/17). Associated findings were hair collar sign, hydrocephaly, spasticity, epilepsy, cleft palate, primary optic nerve atrophy, meningeal arteriovenous fistula, corneal lipodermoid changes, cornea scleralization, nevus flammeus stain, and infantile hemangioma. Fujita et al.[5] reported two cases more, surrounded by a rim of hairs, without bony or neural defects: one case associated with dense dermal melanocytosis and the other with nevus flammeus.

Drolet et al.[3] have proposed that MACC is an abortive form of a neural tube defect. The hair collar sign, regarded as a relatively specific marker for cranial neural tube closure defects, is frequently seen. Histologically, the thin epithelial covering observed resembles that of encephaloceles and meningoceles.[4]

The diagnosis is clinical and a biopsy is not needed. When performed, it shows an atrophic epidermis with loose fibrovascular stroma, edematous dermal stroma, or both.[4] A sonography of the lesion and a transfontanelar sonography should be done to rule out skull defects or cerebral abnormalities.[4] MACC can be visualized on prenatal sonography as a smooth cystic lesion without flow.[1] Conservative therapy is the option of choice.[2]

Dermoscopy may help to rule out other entities (herpes simplex, epidermolysis bullosa, trauma...) since the atrophic epidermis and fibrovascular stroma is evidenced by the hair bulbs and its characteristic translucency (“translucency’s sign”).

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**Conflicts of interest**

There are no conflicts of interest.

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