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CD34 in Human Hair Follicle

To the Editor:

CD34, also known as human hematopoietic progenitor cell antigen, is a heavily glycosylated transmembrane protein expressed on developmentally early lymphohematopoietic stem and progenitor cells, and on a significant number of acute leukemias (Krause *et al*, 1996). In human skin CD34 has been described in vascular endothelial cells, in a subset of dendritic/spindle-shaped cells (Nickoloff, 1991), and in some cutaneous tumors (Cohen *et al*, 1997).

In a previously published study we could demonstrate the expression of CD34 in cells from the outer root sheath of hair follicles (Poblet *et al*, 1994). In that study, sections from normal human skin of formalin-fixed, paraffin-embedded biopsies were immunostained with a monoclonal antiCD34 antibody (clone QBEND/10). A clear membranous staining of outer root sheath cells could be observed. This was the first report to demonstrate the CD34 expression in a certain type of keratinocytes. We showed that the CD34 staining was very specific for epithelial cells of the external root sheath, and that the staining was limited to cells located below the attachment of the arrector pili muscle and above the matrix cells. Because CD34 is a progenitor cell antigen we suggested a possible relation of these cells with stem cells of hair follicles.

We read with interest the excellent article by Trempus *et al* that has recently appeared in this Journal (Trempus *et al*, 2003). These authors demonstrate CD34 immunohistochemical staining of murine hair follicle bulge keratinocytes. In addition they confirmed the presence of CD34 mRNA in that cells. In general, the immunohistochemical staining pattern of murine keratinocytes concords with the staining pattern that we described in human hair follicle keratinocytes, although the bulge region in human hair follicles can not be as readily identified as it is in mice hair follicles.

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Response

To the Editor:

We are writing in response to the Letter to the Editor submitted by Poblet and Jiminez entitled "CD34 in the Human Hair Follicle." In this letter, they point out that we neglected to refer to their 1994 paper "QBEND/10 (Anti-CD34 Antibody) in External Root Sheath Cells and Follicular Tumors" (Poblet

et al, 1994) in our manuscript (Trempus *et al*, 2003) published in a recent issue of JID. We inadvertently omitted that article in our paper and we certainly acknowledge that their findings clearly demonstrated CD34 expression in human follicular keratinocytes and cutaneous tumors. We do not believe that this omission alters the novelty or accuracy of our data and conclusions.

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A Novel *GJB2* (Connexin 26) Mutation, F142L, in a Patient with Unusual Mucocutaneous Findings and Deafness

To the Editor:

Mutations in *GJB2*, encoding the gap junction β-2 protein connexin (Cx) 26, cause hearing loss with or without skin disorders. Mutations in other connexin genes cause hearing loss, skin disorders, peripheral neuropathies, and craniofacial and limb dysmorphisms (**Table I**; Kelsell *et al*, 2001; Rabionet *et al*, 2002; Paznekas *et al*, 2003).

A 2 y old Caucasian female with psoriasiform mucocutaneous involvement, inflammation of mucous membranes, and severe to profound, bilateral, sensorineural hearing loss was ascertained. Hearing loss was diagnosed by auditory brainstem response test (ABR) at 13 mo of age after failure to develop speech. Cutaneous findings include: (1) periorificial and truncal erythematous patches and scaly erythematous plaques that also involve the face, upper extremities, and diaper area (**Fig 1*A***); (2) exuberant granulation tissue around a gastrostomy and on the perianal skin; (3) scaling and obstruction of the external auditory canals; (4) scaly crusted plaques on the scalp; (5) sparse hair that has filled in with age; and, (6) calcinosis cutis of both heels. There is no palmoplantar hyperkeratosis, nail or hair dysplasia. Skin biopsy reveals mostly orthokeratotic hyperkeratosis with focal parakeratosis, acanthosis, and slight papillomatosis of the epidermis, and foci of acute and chronic inflammation in the subepidermal layer and epidermis.

Oral findings include multiple large dental lamina cysts (**Fig 1*B***) that resolved spontaneously, and primary teeth with focal enamel hypoplasia. The oropharyngeal mucosa shows diffuse erythema with focal ulceration, corrugation, and adherent white plaques (**Fig 1*C***). Intermittent angular cheilitis is observed. Histopathologic features of the oral mucosa show hyperparakeratosis, acanthosis, and scattered neutrophils in the superficial epithelium adjacent to an ulcer.

Endoscopy of the upper gastrointestinal tract reveals a denuded and friable esophagus with marked mucosal inflammation inconsistent with Crohn's disease, and esophageal stricture. The mucosal involvement impairs swallowing, resulting in the placement of a gastrostomy.

Other problems include recurrent urinary tract infections, perirectal abscesses, otitis externa, otitis media, iron deficiency anemia, and reactive thrombocytosis. At 17 mo of age, a brain magnetic resonance imaging showed a mild delay in myelination.

Table I. Connexins in deafness, skin disease, and other disorders

Protein	Gene symbol	Phenotypes
Cx26	*GJB2*	Nonsyndromic hearing loss
		Keratitis-ichthyosis-deafness syndrome
		Hystrix-like ichthyosis-deafness syndrome
		Vohwinkel syndrome (mutilating keratoderma and hearing impairment)
		Diffuse palmoplantar keratoderma and deafness
Cx30	*GJB6*	Nonsyndromic hearing loss
		Clouston syndrome (hidrotic ectodermal dysplasia)
Cx30.3	*GJB4*	Erythrokeratodermia variabilis
Cx31	*GJB3*	Nonsyndromic hearing loss
		Erythrokeratodermia variabilis
		Peripheral neuropathy and hearing loss
Cx32	*GJB1*	Charcot-Marie-Tooth disease with or without hearing loss
Cx43	*GJA1*	Nonsyndromic hearing loss
		Oculodentodigital dysplasia

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