The term “congenital ichthyoses” refers to a heterogeneous group of monogenic diseases with gene mutations resulting in a defective skin barrier (1). On the scalp, the scales may be thick and patients may present scarring alopecia, the features of which are not that well known (2–4). There has only been one report of chronic ulceration localized to areas of alopecia to date (5).

**CASE REPORTS**

This paper focuses on 4 new patients. Their characteristics are presented in Table I. All are females presenting very severe and genetically different types of congenital ichthyosis (TGM1 mutations (n = 2), ABCA12 (n = 1), KRT10 (n = 1)). Alopecia presented as denudation centred on the crown (Fig. 1), except in the case of patient 2 who had total alopecia. Interestingly, the ulcerations were localized exclusively in areas of total baldness (Fig. 1). These

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**Table I. Characteristics of the 4 patients in the current case series, and of the case published by Kempton et al. (5)**

| Patient reference or number | Sex | Medical history | Form of ichthyosis/gene involved | Oral acitretin therapy (mg/kg/day) | Age at evaluation (years) | Severity of scales/erythema (0–10) | Severity of ulceration(s) | History of ulceration(s) | Trigger factor | Severity of alopecia | Age at onset of ulceration(s)/duration of follow-up (years) | Severity of ectropion | Global severity of ichthyosis | Baldness | Severity of palmoplantar keratoderma (0–10) | Severity of ichthyosis/gene | Severity of scales/erythema | Number of ulcerations (maximum diameter) |
|----------------------------|-----|-----------------|---------------------------------|-----------------------------------|---------------------------|----------------------------------|---------------------------|--------------------------|----------------|------------------|-----------------------------------------------|---------------------|--------------------------|---------|--------------------------|------------------------|--------------------------|-----------------------------|
| P1                         | F   | –               | Lamellar ichthyosis/TGM1        | –                                 | 20                        | 8/3                              | Stable                     | Stable                   | –              | Moderate         | Very severe/Complete 19/1                             | Very severe         | Very severe              | Incomplete | Very severe/Incomplete 13/7 | Lamellar ichthyosis/ND | Very severe/ND             | 2 (7 cm)                  |
| P2                         | F   | Metastatic breast cancer                      | Lamellar ichthyosis/TGM1        | –                                 | 68                        | 8/8                              | Stable                     | Stable                   | –              | Very severe       | Very severe/Incomplete 4/17                             | Very severe         | Very severe              | Complete   | Very severe/Incomplete 13/7 | Lamellar ichthyosis/ND | Very severe/ND             | 3 (cm)                     |
| P3                         | F   | –               | Ichthyosis with confetti/KRT10  | –                                 | 68                        | 8/10                             | Transient healing/improvement | Transient healing/improvement | Stable       | Very severe       | Very severe/Incomplete 6/10                            | Very severe         | Very severe              | Complete   | Very severe/Incomplete 13/7 | Lamellar ichthyosis/ND | Very severe/ND             | 3 (cm)                     |
| P4                         | F   | –               | Harlequin ichthyosis/ABCA12     | –                                 | –                         | 6/10                             | Stable                     | Stable                   | –              | Very severe       | Very severe/Incomplete 13/7                             | Very severe         | Very severe              | Complete   | Very severe/Incomplete 13/7 | Lamellar ichthyosis/ND | Very severe/ND             | 1 (4 cm)                   |
| Kempton et al., 2018 (5)   | M   | –               | Lamellar ichthyosis/ND          | –                                 | –                         | –                               | –                          | –                        | –              | –                | –                                                             | –                   | –                        | –         | –                        | –                       | –                        | –                            |
areas were atrophic and covered by erythematous patches, small erosions and crusts. The hairy scalp was covered with scales. All patients reported intermittent surrounding pustular lesions. The ulceration(s) had appeared during childhood or adulthood, with no apparent trigger factor, and were chronic with transient periods of improvement. All patients received long-term oral acitretin therapy from the outset and wore a wig/scarf (without hairpins). Bacterial swabs revealed abundant *Staphylococcus aureus* in 2 cases. Histological examination of the ulcerations confirmed non-specific inflammatory ulceration (ulcerated epidermis replaced by fibrin and leukocyte exudate, underlying oedematous dermis with numerous capillaries, inflammatory infiltrate [lymphocytes, histiocytes, plasmocytes and polymorphonuclear neutrophils]). A second biopsy taken from patient 2 also revealed *in situ* squamous cell carcinoma. Topical or intralesional steroids improved the ulceration, but did not promote complete recovery.

**DISCUSSION**

The only patient reported with a similar lesion was a young man presenting lamellar ichthyosis, who was treated successfully with dehydrated human amnion/chorion membrane allograft (5). The author concluded the patient had erosive pustular dermatosis of the scalp (EPDS), a rare entity clinically characterized by chronic eruption of scalp pustules, erosions, crusts and scarring alopecia. Although EPDS has occasionally been reported in children, it mostly affects older individuals with androgenetic alopecia and actinic damage of the scalp, and is often preceded by trauma (6). The characteristics of our 4 patients do not closely match the diagnosis of EPDS (2 children and a young woman, no sun exposure as they avoided the sun and wore a wig/scarf, no reported trauma). The cause of such ulceration is unknown. Staphylococcal growth may represent secondary colonization. These ulcerations may be linked to skin barrier anomalies responsible for abnormal inflammatory processes and disturbed microflora. Gene mutations may also be responsible for the selective expression of antigenic proteins by hair follicles (3). Other contributory factors cannot be ruled out: repeated trauma/maceration secondary to wearing a wig or scarf and pruritus. Scalp ulcerations might also be an unusual and uncommon side-effect of long-term retinoid therapy.

The presence of *in situ* squamous cell carcinoma in one patient raises questions. It may be due to the skin-ageing process (despite long-term sun avoidance) but could also be indicative of the potentially cancerous nature of these chronic ulcerations. Close follow-up is therefore required.

In conclusion, these new cases increase our knowledge of alopecia-related scalp anomalies, but further studies are required in order to gain a better understanding of this rare condition and identify new treatments.

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