Localized scleroderma (LS) in childhood is a relatively rare fibrosing disorder of the skin and underlying tissues. Early subtle skin changes include hyperpigmented, hypopigmented, erythematous, or purplish patches. In particular, *En coup de sabre* (ECDS), a linear type of LS located on the scalp and/or forehead, may lead to cosmetic deformities, sometimes including the skull and neurological complications (e.g. seizures). Thus, early diagnosis and treatment of this condition is important. However, rare cases of early LS that mimicked and were misdiagnosed with an acquired port-wine stain (PWS), a capillary malformation, have been reported recently (1–7). We report here 2 such cases along with a literature review.

**CASE REPORTS**

**Patient 1.** A 3.5-year-old Japanese girl presented with erythematous plaques and pruritus on her right mandibular area for more than 6 months (Fig. 1a). Histopathological findings included thickening and homogenization of collagen bundles, dilation of capillaries and perivascular lymphocytic infiltrates. Lymphocyte and plasma cell infiltrates were also found at the junction of the septa and lobules (Fig. 1b). Although her skin lesions first suggested a diagnosis of PWS, we finally diagnosed her with linear scleroderma. She was treated with oral betamethasone for 6 months and her erythema receded completely, leaving residual indurated plaques along her right mandible (Fig. 1c). Despite subsequent treatment with tranilast for 2 years, a depression with skin atrophy occurred (Fig. 1d).

**Patient 2.** A 2-year and 11-month-old Japanese girl presented with a light brownish sclerotic linear plaque on the right side of her face and head, involving the eyelid, forehead and parietofrontal scalp, with alopecia and bony atrophy (Fig. 1e). Her mother first noticed redness on the area at the age of 15 months, and she was diagnosed with PWS by a dermatologist at the age of 20 months (Fig. 1f). A skin biopsy showed thickening and homogenization of collagen bundles and perivascular lymphocytic infiltrate in the subcutaneous tissues as well as in the dermis. She was diagnosed with ECDS. A computed tomography scan showed a curved depression of the skull, while...
an electroencephalogram was normal. Oral prednisolone for a period of 6 months improved the sclerosis, but not the scar-like appearance. In both patients, results of blood biochemical examinations were normal and an anti-nuclear antibody test was negative. No trauma or other relevant medical or family histories were reported.

DISCUSSION

Fifteen cases of LS initially misdiagnosed as PWS or salmon patch, including the 2 cases presented here, have been reported in the literature (1–7), and 11 cases described in detail were summarized (Table SI1). The female: male ratio was 10:1. Except for a single case of a 24-year-old Korean woman, the remaining 10 cases were children, including one congenital case, with a mean age at disease onset of 2.6 years (range 0–6 years; median 2.0 years) and a time from onset to LS diagnosis of 0.4–4 years. The clinical subtype was linear scleroderma in all the cases except for case 6, which had insufficient clinical information. The affected sites were the face and/or scalp in 10 cases, except for one with limb involvement, while extracutaneous manifestations, such as central nervous system involvement, were not described. No significant triggers were reported in any of the cases, but 4 cases were treated with pulsed dye laser (PDL) because they were misdiagnosed as PWS. As trauma is considered as a candidate trigger for LS, PDL may have triggered or accelerated the development of LS. However, case 2 treated with PDL later developed scleroderma even on an area not treated with PDL, thus we do not think PDL is a trigger. A negative anti-nuclear antibody result was described in 4 cases examined.

In the review of 750 cases of juvenile LS, the mean age at disease onset was 7.3 years (range 0–16 years), female: male ratio 2.4:1, and linear scleroderma, including ECDS and progressive hemifacial atrophy, was the most frequent subtype (65%), but the face and/or scalp presentation of the linear type accounted for only 15% of the total (8). In another review of 136 cases of juvenile LS, the mean age at disease onset was 8.2 years (median 8.1 years; range 0–18 years), female: male ratio 2.6:1, and linear scleroderma was the most frequent subtype (51%), but the face and/or scalp presentation accounted for only 20% (9). Thus, LS initially presenting with PWS-like appearance has a unique tendency to occur in very young children, being most common in girls, and mostly located on the face and/or scalp as a linear subtype. The early stage of LS has been known to be an inflammatory stage with lymphocytic infiltration, oedema, and increased vascularity observed on histopathological examination (10). Inflammation involving T-helper cells and their associated cytokines may be pivotal for initiating the abnormal fibrosis (11). Thus, some lesions could be recognized by the presence of an erythematous or violaceous lesion mimicking PWS. Interestingly, in a review of 50 children with LS, the first doctor misdiagnosed approximately one-third of them with inflammatory diseases, such as atopic dermatitis (5). In a review of 6 congenital LS patients, 2 were initially misdiagnosed with a skin infection and 1 with a salmon patch (case 1), suggesting that inflammatory erythema may be a more common presentation in early childhood. The skin of the face and neck is the most susceptible area for vascular anomalies including PWS (12) and infantile haemangioma (13), which may explain the high frequency in these areas in the 11 cases reviewed here. As acquired PWS is very rare, LS must be suspected in infants presenting with a PWS-like lesion on the face or scalp.

The authors declare no conflicts of interest.

REFERENCES