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P018 Puzzling vesicles: a clinicopathological challenge

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An 8-year-old male child presented with a history of multiple vesicles and bullae on the ear, nose, upper trunk, lower legs and feet for the past 4 years. There was no history of photosensitivity. There was no history of similar conditions in family members. On examination, the child had erosions with haemorrhagic crusting on the above-mentioned sites. The lesions were healing with hypopigmentation and scarring. A few milia-like lesions were seen on the nose and ears. Superficial tiny erosions with haemorrhagic crusting were
present on the lip mucosa and tongue. Dystrophy of the left great toe was also noted. Differential diagnoses of Kindler syndrome, porphyria cutanea tarda, bullous pemphigoid and chronic bullous dermatosis of childhood were considered. A 3-mm punch biopsy from the vesicle revealed epidermis with psoriasiform hyperplasia and hyperkeratosis. The upper dermis showed a perivascular and perifollicular lymphomononuclear infiltrate with multiple eosinophils. A perilesional 3-mm punch biopsy was sent for direct immunofluorescence, which revealed at least three IgG linear deposits along the dermoepidermal junction in an N-serration pattern. There were no IgA or IgM deposits. A genetic analysis showed no mutation in the FERMT1 gene. Porphyrin levels in the blood, urine and stool were normal. Based on the histopathology and direct immunofluorescence findings, the patient was diagnosed with childhood bullous pemphigoid. Weight-based azathioprine was started and there was improvement in the lesions within 2 months of initiation of therapy. Bullous pemphigoid is a subepidermal blistering disorder that commonly affects older patients (60–80 years), but it can rarely affect children. It demonstrates considerable clinical and histopathological overlap with other acquired and/or congenital blistering disorders. A high index of suspicion coupled with appropriate investigations are imperative for diagnosis. Histopathology and direct immunofluorescence are more cost-effective. In a limited-resource setting, this can eliminate the need for more expensive tests, such as genetic analysis.