

Figure 3. The epithelial lining is composed of three cell types. The insets show cells with mucinous differentiation (m), squamoid cells with eosinophilic cytoplasm (e), and polygonal cells with clear cytoplasm (c). Most cells had regular, round, small nuclei. Some focal areas of increased mitotic rate and nuclear atypia are not shown (HE, original magnification: $\times 20$, scale bar: $200 \mu\text{m}$, insets: $\times 40$, image width $100 \mu\text{m}$).

approximately 10 instances of MEC at *any* primary site in children as young as our patient (7 years) have been reported (3).

Histologically, MEC is characterized by three cell types: mucinous, squamous, and clear (4). Lesions are typically well-circumscribed and demonstrate peritumoral fibrosis, variability in the epithelial lining, mucinous, and papillary features (2,5). Immunohistochemical analysis reveals positive staining for cytokeratin and carcinoembryonic antigen. Histologic grade correlates well with clinical behavior, independent of primary tumor location, and low-grade MEC rarely metastasizes (2,6). Local recurrences may occur after incomplete excision (6,7).

Clinical diagnosis of MEC of the skin is challenging (2). First, MEC of the skin may be primary or metastatic. Whereas primary MEC of the skin tends to be low-grade, metastatic skin MEC tends to be high-grade and is considered to indicate a poor prognosis. A recent review (2) identified only eight well-documented occurrences of primary MEC of the skin (two deaths) and three of MEC metastatic to the skin. Secondly, MEC of the skin is commonly confused with adenosquamous carcinoma (ASC) of the skin (5), which may present at various pri-

mary sites, including the cervix, lungs, intestines, pancreas, throat, and skin. This entity is characterized histologically as a more high-grade, aggressive malignancy with areas of both squamous cell carcinoma and adenocarcinoma. It is less circumscribed, and does not demonstrate peritumoral fibrosis, variability in the epithelial lining, mucinous, or papillary features (2,5). Additionally, MEC is dermal-based whereas ASC may be intraepidermal (2,5).

In summary, to our knowledge, this is the first report of primary MEC of the skin occurring in childhood. Primary MEC of the skin should be distinguished from both MEC metastatic to the skin and ASC, given the important morphologic and prognostic differences.

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LICHEN PLANUS APPEARING SUBSEQUENT TO GENERALIZED LICHEN NITIDUS IN A CHILD

To the Editor:

Lichen nitidus (LN) is an uncommon inflammatory skin disease, primarily of children (1). It is distinguished

from lichen planus (LP) on both clinical and histologic grounds. However, the possibility that LN represents a micropapular variant of LP has never been definitively excluded (2). We report a child who developed generalized LN which was soon followed by LP.

A 7-year-old boy in good health was seen with a 1-year history of multiple, asymptomatic skin papules. Skin examination showed monomorphous, pinpoint, round, or polygonal shiny papules scattered on his shoulders, limbs, and trunk. Histologic examination showed circumscribed nests of lymphocytes and histiocytes and occasionally epithelioid and Langhans cells in the upper dermis. The rete ridges on the margins were elongated, creating the image of a claw clutching a ball. The clinical diagnosis of LN was confirmed. Due to the lack of symptoms, no treatment was suggested. Two months later, while LN lesions remained stationary, the patient developed a pruritic, symmetrically distributed eruption of violaceous papules isolated, grouped or sometimes coalescing in plaques on his limbs, wrists, trunk, and genital area (Fig. 1A,B). Clinical and histopathologic findings of the latter lesions were typical of LP. Biochemical investigations did not disclose any abnormality, in particular no infection with the hepatitis C virus was found. Narrow band-UVB phototherapy was performed with good results on the LP lesions which regressed quickly with residual hyperpigmentation. Remission of the skin manifestations of LP occurred after 2 months of treatment. At a follow-up of 12 months the boy is free from skin lesions of LP. Some residual lesions of LN are still present in some circumscribed areas of the trunk.

Lichen nitidus is a rare disorder of unknown etiology which clinically presents with dome shaped, shiny, asymptomatic papules, 2–5 mm in diameter. Solitary lesions, or less frequently, generalized eruptions and linear distribution have been observed. It has been proposed that immune alterations can be associated with this entity, particularly the generalized form (1).

The association with LP is still a subject of debate. Clinically LN is distinct from LP, as it is neither pruritic nor violaceous and is characterized by small, monomorphous papules. In addition, a distinctive histopathology profile is observed. However, similar histologic findings in early lesions of both diseases have been reported. In particular epithelioid and giant cells, which are considered typical features of LN, have been described in the skin or nail lesions of LP (3–5). A review of the literature reveals that LN may accompany clinical variants of LP and both conditions may occur together in the same patient (2). In 1988, Aram (6) reported a patient with associated of LP and LN who had a beneficial response to treatment with etretinate. In 1995 Kano et al (7) reported a patient with a 2-year history of Crohn disease who developed both LP and LN in addition to erythema nodosum lesions. In the same year, Kawakami and Sama (8) reported generalized LN appearing subsequent to LP in a 27-year-old man.

Of interest, ultrastructural changes of LN were found identical with those reported in LP (9–10), as well as immunophenotypic findings of a marked excess of CD4+ cells over CD8+ cells and a large number of

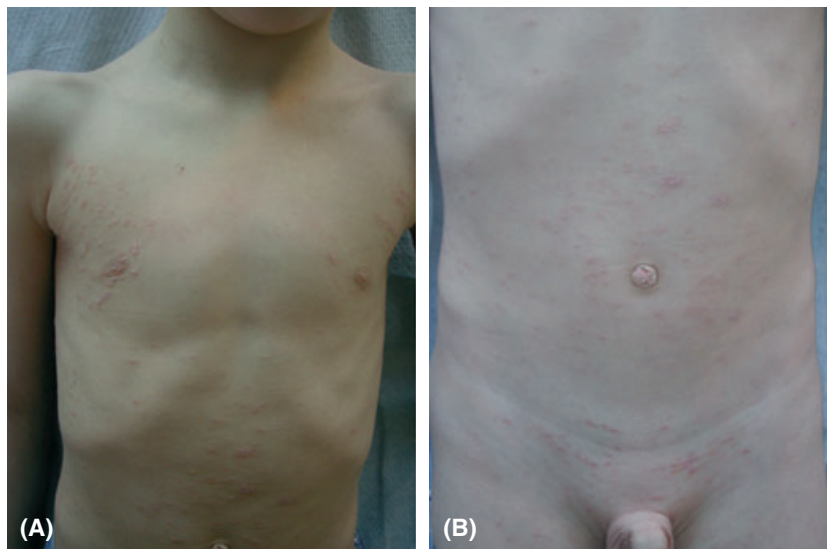


Figure 1. (A) Multiple violaceous papules, often coalescing into plaques with fine linear white scales, grouped on the pectoral area, axillae and pubic area, and (B) diffusely dispersed on the abdomen.

CD1+ cells (11). However the presence of a more consistent quota of helper T-cells in LP lesions and a more heterogenous infiltrate in LN was pointed out in another study (12).

To the best of our knowledge, this is the first instance of an association of LP and LN in a child. As generalized LN is a rare condition, as is childhood LP, it is not likely that the coexistence of the two diseases in our patient was a fortuitous one. This further occurrence of an association of LN and LP does support the view that both conditions could be precipitated by a common unidentified etiologic factor. Thus, LP and LN could represent different, but closely related diseases with possible similar etiologic factors priming different immunologic mechanisms.

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METHYLMALONIC ACIDEMIA PRESENTING WITH AN ICHTHYOSIS VULGARIS-LIKE ASPECT

To the Editor:

Methylmalonic acidemia (MMA) is a rare inborn error of propionate metabolism, inherited in an autosomal recessive fashion and genetically heterogeneous. It is produced by the diminished activity of methylmalonyl CoA mutase or its coenzyme adenosylcobalamine (vitamin B12) that affects the normal catabolism of four essential amino acids (valine, isoleucine, threonine, and methionine) (1,2). Onset of the disease occurs soon after birth and consists of a severe, progressive acidosis metabolic state. Principal signs are failure to thrive, poor feeding, vomiting, hypotonia, convulsions, and mental retardation. Conventional treatment includes dietary protein restriction, bicarbonate, carnitine, and metronidazole. The course of MMA presents frequent complications and sometimes a fatal outcome (3,4).

Cutaneous manifestations associated with MMA are infrequent and not well recognized. It is still not clear enough whether skin lesions are related to the specific enzyme deficiency or are the result of the therapeutic dietary amino acid restrictions (3). Some authors classified skin findings in MMA based on their clinical appearance, into five types: superficial scalded skin, superficial desquamation (the most typical lesions, which mainly appear after metabolic decompensation), bilateral and periorificial dermatitis (the most frequent MMA skin manifestation, which resembles acrodermatitis enteropathica), psoriasiform lesions, and alopecia (1).

We report a 2-year-old boy, born to nonconsanguineous parents, who was diagnosed as having MMA at 21 months of age. He was referred to our department due to a persistent ichthyosiform state since birth, associated with mild pruritus. Family history was unremarkable for cutaneous diseases. Physical examination found diffuse desquamation without associated erythema that only spared the face and flexural folds. Light-gray scales covered the trunk and upper extremities (Fig. 1) while more accentuated and adherent brownish scales were present on the anterior aspects of the legs. The presumed clinical diagnosis was ichthyosis vulgaris and a skin punch biopsy from the chest was performed. The main histologic findings were consistent with orthohyperkeratosis alternating with small foci of parakeratosis, elongated rete-papillae, and a preserved granular layer (Fig. 2). Therefore, considering the congenital presentation of our patient's cutaneous disease together with the absence of familial antecedents of ichthyosis vulgaris, the negativity for personal and familiar atopy, and the histologic feature of a conserved granular layer in the skin