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Images

Acrodermatitis dysmetabolica in phenylketonuria

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A 17-month-old girl with a history of phenylketonuria (PKU) and chronic kidney disease (CKD) secondary to unilateral renal agenesis presented with failure to thrive, diarrhea, and an extensive skin rash. Her parents reported increased irritability, decreased oral intake, and a gradually worsening rash over the last 14 days, with watery diarrhea present for 3 days. The patient was on a special amino-acid-restricted diet formula since birth for PKU and CKD. On examination, a diffuse erythematous confluent rash was present in the perioral region and the inguinal and perianal areas (as shown in Figs. 1 and 2). Her phenylalanine, tyrosine, and branched-chain amino acid contents were very low on the amino acid quantitative panel. Zinc levels and kidney function tests were normal. The patient's condition was clinically diagnosed as acrodermatitis dysmetabolica, which was confirmed by a skin biopsy. Skin findings such as acrodermatitis enteropathica are common in patients with aminoacidopathies, including PKU, and are also found in subjects with organic acidemias, urea cycle disorders, and essential fatty acid deficiency.[1] However, unlike acrodermatitis enteropathica, the rash in acrodermatitis dysmetabolica usually does not respond to zinc supplementation. The etiology of the rash is secondary to essential amino acid deficiency, leading to keratinocyte growth arrest.[2] The treatment involves timely supplementation of the amino acids that are deficient. Frequent monitoring of amino acid levels and avoidance of amino acid imbalance are key factors in the prevention of this skin condition. The patient was started on a special diet with sufficient amino acids, and her rash gradually improved with dietary adjustments and emollients.

Figure 1 Acral distribution of the rash—face.

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Figure 2 Acral distribution of the rash—groin.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

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Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.pedneo.2018.05.005.