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# Pediatric Dermatology WILEY

# A sharply marginated erythematous dermatitis in a toddler

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# **CASE PRESENTATION**

A 33-month-old boy was referred for a 2-month history of a diffuse rash, unsuccessfully treated with oral dexamethasone and fluconazole. He was delivered at term with normal birthweight. His immunizations were up-to-date. According to his parents, he recently began to lose previously acquired motor and verbal skills. There was no family history of rheumatologic or hematologic diseases. On admission, the child was irritable, non-verbal, but had normal vital signs. Physical exam was remarkable for mild hepatomegaly and an inability to walk. Skin examination showed hair depigmentation (Figure 1); a sharply marginated erythematous dermatitis with irregular raised borders on the limbs, trunk, and perianal region (Figures 2 and 3), and pitting edema in both feet (Figure 3).

# WHAT IS THE DIAGNOSIS?





### **FIGURE 1**

**FIGURE 2** 



**FIGURE 3** 

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### **Diagnosis: Kwashiorkor**

Laboratory analysis documented severe hypoalbuminemia, irondeficiency anemia, and hypogammaglobulinemia. Coagulation workup showed prolonged prothrombin time secondary to deficiency of vitamin K dependent proteins. Urinalysis, platelet count, acute phase reactants, and blood zinc level were within normal limits. Dietary history revealed that since age 24 months, he was exclusively consuming rice milk because of a presumed food allergy.

The diagnosis of kwashiorkor was confirmed by the response to a therapeutic diet, which initially consisted of peptide-based enteral feeds, then shifted to an oral milk-based formula, and finally transitioned to a full diet. Intramuscular vitamin K and a daily multivitamin supplement were also administered. Within a few weeks, he exhibited marked clinical improvement of skin lesions and psychomotor state.

# DISCUSSION

Kwashiorkor is a form of malnutrition related to insufficient protein intake. leading to hypoalbuminemia and low serum levels of essential nutrients.<sup>1</sup> Major cutaneous features include edema, a characteristic dermatosis and hair abnormalities. The dermatosis is characterized by erythematous lesions with varying degrees of desquamation and erosion. The edges of these lesions look like paint lifting up and about to peel off, a pattern described as "flaky paint dermatosis."<sup>2</sup> Proteindeficiency may disturb the normal cycling of the hair follicles and cause dysplastic or atrophic anagen roots with increased proportion of hairs in telogen, a striking reduction in root bulb diameter, incomplete outer sheaths, and a reduced melanin biosynthesis resulting in bulb dyspigmentation.<sup>3</sup> Thus, visible changes to the hair shaft can include dry, lusterless, fine, unruly, and brittle hair; hair loss and hypochromotrichia. Our patient displayed the typical horizontal pigmented bands developing within the hair ('flag sign'), due to initiation of melanin deposition during times of improved nutrition. Other findings include hepatomegaly and neurodevelopmental impairment, leading to motor, cognitive, and socioemotional alterations.<sup>4</sup>

Child malnutrition is a global issue, declining worldwide except in Africa.<sup>5</sup> In developed countries this condition is mostly associated with malabsorption-related diseases (e.g., cystic fibrosis, Crohn's disease), but it can also occur in the context of increasingly prevalent dietary issues such as food fads, food allergy with elimination diets, and nutritional ignorance.<sup>6,7</sup> In our case, the dietary history was crucial to diagnosing kwashiorkor. Notably, rice milk contains a negligible amount of protein (1.7 g/L) compared with the protein content of cow's milk (33 g/L).<sup>8</sup> Several cases of kwashiorkor due to elimination diets with inadequate nutrients have been reported, highlighting the risks associated with a self or parent-initiated restrictive diet without medical supervision.7

Differential diagnosis of the eruption includes atopic dermatitis, vasculitis, infections, and other nutritional disorders.<sup>8</sup> The generalized distribution of the eczematous lesions in atopic dermatitis may

resemble kwashiorkor, which could be unrecognized in overlapping cases. Vasculitis can result in a wide range of multi-organ and skin manifestation, including petechiae, purpura, nodules, livedo, urticaria, ulcers, and alopecia.<sup>9</sup> Henoch-Schonlein purpura (HSP), the most common vasculitis in children, may display a painful edema in both feet mimicking kwashiorkor. However, HSP is characterized by a palpable purpuric rash predominantly on lower limbs and buttocks, and by other systemic symptoms, including gastrointestinal, joint, and kidney involvement.<sup>9</sup> Among nutritional disorders, dermatitis resembling that of kwashiorkor has been described in patients with inherited or acquired zinc and biotin deficiencies. Moreover, biotin and other B vitamins deficiencies usually display neurological signs.<sup>8</sup> As children with kwashiorkor may have a concomitant multivitamin deficiency, the presence of edema is helpful in distinguishing kwashiorkor from other nutritional disorders. Whereas in kwashiorkor the caloric intake is adequate, in marasmus there is deficiency of both protein and calories, resulting in emaciation with dry and scaling skin in the absence of edema.<sup>1</sup> Complications of proteinenergy malnutrition include hypothermia, hypoglycemia, encephalopathy, heart failure, and infections.

Alopecia, as might be expected, may occur in all nutritional disorders, but dyspigmentation typically appears in kwashiorkor, marasmus, and essential fatty acid deficiency.<sup>3</sup>

Management of kwashiorkor starts with correction of metabolic abnormalities. A nutrition consult is recommended to set up dietary therapy and prevent refeeding syndrome. Milk-based formulas are the treatment of choice along with a multivitamin supplement.<sup>10</sup> Recovery of brain function is plausible if the nutritional status is re-established while the affected growing process is still occurring.<sup>4</sup>

This case highlights the importance of a detailed dietary history and careful examination of skin and appendages in evaluating patients with dermatoses suspicious for nutritional deficiency.

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