Acute Kwashiorkor in the Setting of Cerebral Palsy and Pancreatic Insufficiency

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PRACTICE **POINTS**

- Pancreatic exocrine deficiency, confirmed by low stool elastase, can lead to kwashiorkor and requires a high index of suspicion for diagnosis.
- Kwashiorkor is not only seen in developing countries but also in certain at-risk populations in economically advantaged countries.
- For multiple reasons, patients with cerebral palsy are at particular risk for nutritional deficiencies including kwashiorkor.

To the Editor:

Kwashiorkor, or protein-calorie malnutrition, is a common issue in developing countries subject to starvation. In economically advanced nations, however, kwashiorkor is extremely rare and may appear in children placed on restrictive diets instituted by well-meaning guardians. Kwashiorkor also may occur because of gastrointestinal malabsorption. We present a unique case of kwashiorkor that revealed an underlying diagnosis of pancreatic insufficiency.

A 12-year-old girl presented to the hospital with 4 days of watery nonbloody diarrhea occurring with every feeding as well as new onset of presumed diaper dermatitis that had not responded to nystatin cream. Facial swelling also was noted the day prior to admission. Her medical history was notable for cerebral palsy secondary to nonaccidental trauma, leaving the patient nonverbal and quadriplegic. She had numerous prior admissions for sepsis with marked hypotension and more recently was diagnosed with insulin-dependent type 2 diabetes mellitus. She had never lived outside the United States and resided at home with her adoptive parents.

Physical examination revealed a nonverbal underweight girl (weight, 25 kg). Large areas of denudation with surrounding desquamated skin resembling flaking enamel paint covered the buttocks and posterior legs bilaterally (Figure). She had linear hyperpigmented patches on the dorsal hands with one superficial erosion on the left wrist. Marked periorbital edema as well as nonpitting edema of the face, arms, and legs were present.

Upon additional questioning, the patient's adoptive parent reported a diet of formula containing 1.0 cal/mL with 200-mL feedings 3 times daily through a Geiger-Müller tube, providing a daily protein intake of approximately 17.7 g per day (0.7 g/kg per day). On the day of admission, abnormal laboratory findings included low protein and albumin levels at 4.6 g/dL (reference range, 5.7–8.2 g/dL) and 2.1 g/dL (reference range, 3.2–4.8 g/dL), respectively; an elevated aspartate aminotransferase level of 73 U/L (reference range, 10–34 U/L); and an elevated alanine aminotransferase level of 80 U/L (reference range, 10–40 U/L). Based on the patient's characteristic clinical findings and abnormal laboratory values, a diagnosis of acute kwashiorkor was made. Although the zinc level was low at 0.29 μg/mL (reference range, 0.66–1.10 μg/mL), the patient did not have any periorificial involvement to support a diagnosis of acrodermatitis enteropathica.

Upon further workup, stool elastase was measured at less than 50 μg per gram of stool (reference range, >200 μg pancreatic elastase per gram of stool), confirming a diagnosis of severe pancreatic insufficiency. Pancreatic enzyme supplementation was initiated along with an increase in protein intake to 1.5 g/kg per day. The patient's hospital course was complicated by respiratory distress and sepsis, leading to a prolonged hospital stay. A component of refeeding syndrome may have contributed to the patient's respiratory distress.

Kwashiorkor, a form of protein malnutrition, is caused by inadequate protein intake and usually is seen in

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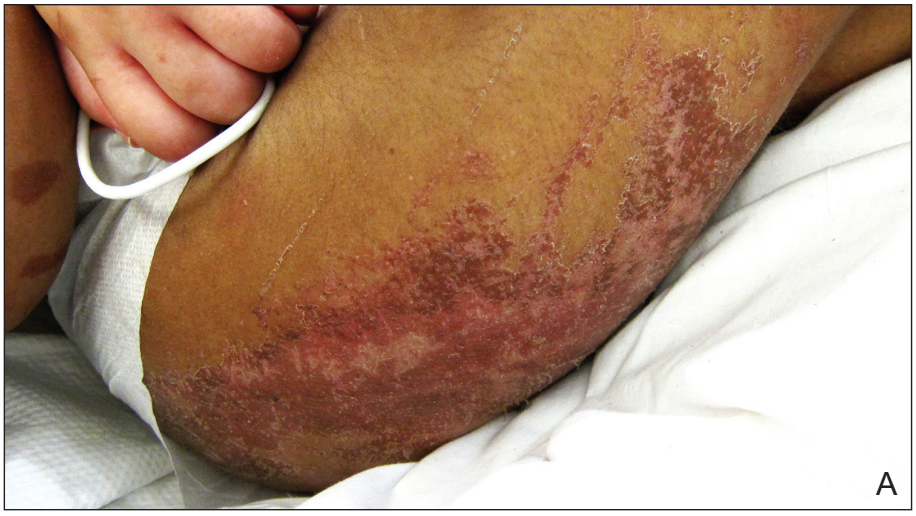

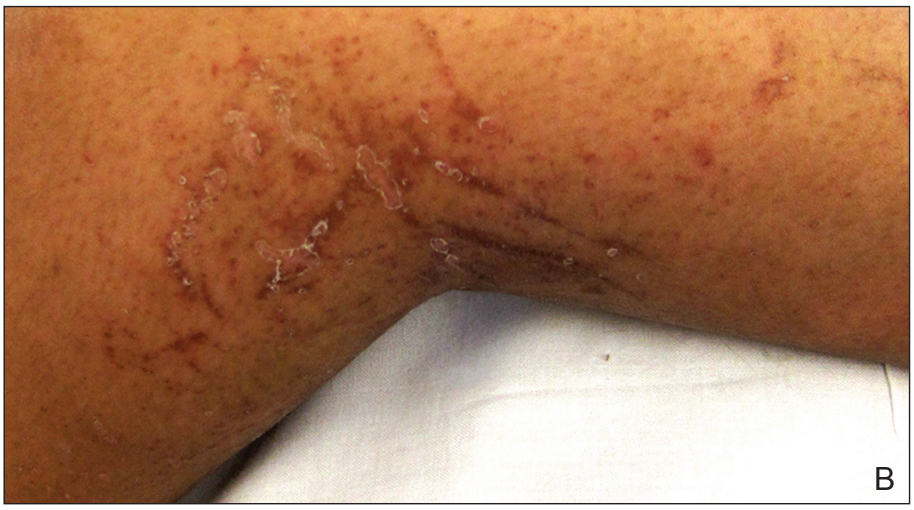

A, The right buttocks and posterior thigh showed patchy reddish brown discoloration with marked exfoliation and areas of hypopigmentation, giving an appearance of flaking enamel paint. There also was notable edema of the patient's fingers. B, The lateral right lower extremity showed areas of hyperpigmented desquamation in a linear configuration.

developing countries when children are weaned from breastmilk to a diet high in starch and low in protein. It is characterized by edema, growth retardation, a characteristic dermatosis, depigmentation of hair, lethargy, and irritability.[1] If left untreated, kwashiorkor can be fatal. Skin changes associated with kwashiorkor first occur in areas of friction or pressure. The skin develops patches of hyperpigmentation that subsequently desquamate in a pattern likened to flaky paint. In the current case of a nonmobile child with diarrhea, prominent involvement of the buttocks and thighs would be expected. This dermatosis does not appear in marasmus and is pathognomonic for kwashiorkor when seen in a child with edema.[2]

Children in the United States developing kwashiorkor secondary to severely restrictive diets has been reported.[3] However, kwashiorkor also may occur due to underlying chronic malabsorptive disease. There have been rare reports of children with cystic fibrosis presenting with kwashiorkor,[4] as well as a case of kwashiorkor secondary to underlying infantile Crohn disease.[5]

Cerebral palsy is associated with multiple different risk factors for malnutrition. Musculoskeletal deformities, oral-motor difficulties, medication side effects, limited communication skills, compromised pulmonary status, and poor muscle tone can all contribute to energy and nutrient deprivation.[6] A 2018 study including 728 children registered into the Bangladesh Cerebral Palsy Register between January 2015 and December 2016 demonstrated that more than two-thirds were underweight (70.0%) and stunted (73.1%) and that children with tri/quadriplegic cerebral palsy presented with the highest proportion of severe malnutrition.[7] In another report (N=142), up to 85% of children with spastic quadriplegia had severe feeding problems,[8] making this population particularly high risk for poor nutritional status.

Pancreatic exocrine insufficiency is characterized by reduced secretion of amylase, lipase, and protease, and it may result in diarrhea, weight loss, malabsorption of essential nutrients, and malnutrition. Pancreatic exocrine insufficiency may occur in the setting of chronic

pancreatitis, pancreatic surgery, and cystic fibrosis. Our patient had numerous hospitalizations for sepsis marked by hypotension, and in the absence of more typical causes, we postulate that both endocrine and exocrine pancreatic damage resulted from prolonged hypotension. A sweat chloride test was not performed, as the patient had not experienced frequent pulmonary infections or other signs of cystic fibrosis.

According to a report from the Food and Agriculture Organization of the United Nations/World Health Organization/United Nations University (FAO/WHO/UNU), protein should provide at least 10% of the total caloric intake in a child.[9] Although the adoptive parent approximated that our patient received 12% of her daily calories in the form of protein, the amount that she absorbed in the context of pancreatic insufficiency was undoubtedly much lower.

In this case, the diagnosis of kwashiorkor led to the discovery of underlying pancreatic exocrine insufficiency. Low stool elastase confirmed the diagnosis. Because kwashiorkor is rare in developed countries, the classic signs and symptoms may go unrecognized, which can lead to delayed diagnosis and notable morbidity and mortality. New-onset edema and desquamative rash in a child, especially a child with cerebral palsy, should alert physicians to the possibility of acute kwashiorkor and prompt investigation into underlying medical issues that may have contributed to its development.

REFERENCES

1. Trowell HC, Davies JN, Dean RF. Kwashiorkor. II. clinical picture, pathology, and differential diagnosis. *Br Med J.* 1952;2:798-801.
2. Latham MC. The dermatosis of kwashiorkor in young children. *Semin Dermatol.* 1991;10:270-272.
3. Liu T, Howard RM, Mancini AJ, et al. Kwashiorkor in the United States: fad diets, perceived and true milk allergy, and nutritional ignorance. *Arch Dermatol.* 2001;137:630-636.
4. Phillips RJ, Crock CM, Dillon MJ, et al. Cystic fibrosis presenting as kwashiorkor with florid skin rash. *Arch Dis Childhood*. 1993; 69:446-448.
5. Al-Mubarak L, Al-Khenaizan S, Al Goufi T. Cutaneous presentation of kwashiorkor due to infantile Crohn's disease. *Eur J Pediatr.* 2010;169:117-119.
6. Wittenbrook W. Nutritional assessment and intervention in cerebral palsy. *Practical Gastroenterol*. Feb 2011;92:16-32. http://www.practicalgastro.com/pdf/February11/WittenbrookArticle.pdf.
7. Jahan I, Muhit M, Karim T, et al. What makes children with cerebral palsy vulnerable to malnutrition? findings from the Bangladesh cerebral palsy register (BCPR)[published online April 16, 2018]. *Disabil Rehabil*. doi:10.1080/09638288.2018.1461260.
8. Stallings VA, Charney EB, Davies JC, et al. Nutrition-related growth failure of children with quadriplegic cerebral palsy. *Dev Med Child Neurol*. 1993;35:126-138.
9. World Health Organization. *Energy and Protein Requirements: Report of a Joint FAO/WHO/UNU Expert Consultation.* Geneva, Switzerland: World Health Organization; 1985. Technical Report Series 724.