Kwashiorkor and Concurrent Acquired Acrodermatitis Enteropathica in a Case with Hirschsprung’s Disease
-A Case Report and Review of the Literature

Yi-Shan Liu1 Pei-Yu Lo1 Tien-Yi Tzung1, 3 I-Fei Huang2

Kwashiorkor is a form of protein-energy malnutrition with a characteristic “flaky paint” dermatosis. Patients with kwashiorkor may have a low serum level of zinc. However, most patients fail to show the typical cutaneous manifestation of zinc deficiency, namely, acrodermatitis enteropathica. We herein reported a patient with concurrent kwashiorkor and acquired acrodermatitis enteropathica. This 2-year-and-8-month-old boy, a case of Hirschsprung’s disease, was admitted due to enterocolitis. Ten days after admission, generalized hyperkeratotic and hyperpigmented plaques with symmetrical scaly and erosive eczematous lesions around the body orifices were observed. Laboratory data revealed hypoalbuminemia (1.7 g/dL) and hypozincemia. After intravenous administration of human albumin together with zinc gluconate 5 mg once a day, his skin rash subsided gradually in ten days. (Dermatol Sinica 25: 43-47, 2007)

Key words: Kwashiorkor, Acrodermatitis enteropathica, Hirschsprung’s disease, Zinc deficiency

From the Departments of Dermatology1 and Pediatrics,2 Veterans General Hospital-Kaohsiung and the Department of Dermatology, National Yang-Ming University, Taiwan3

Accepted for publication: September 21, 2006
Reprint requests: Pei-Yu Lo, M.D., Department of Dermatology, Veterans General Hospital-Kaohsiung, No 386, Ta-Chiung 1st Rd, Kaohsiung, Taiwan.
TEL: 886-7-3422121 ext. 4304  FAX: 886-7-3468209  E-mail: pylo@vghks.gov.tw
INTRODUCTION

Kwashiorkor, caused by insufficient protein intake, is endemic in Africa and in developing countries. However, it may occur in developed countries associated with various malabsorptive and malnutritional conditions. Acquired acrodermatitis enteropathica is an unusual disorder caused by a deficiency of zinc. Coexistence of kwashiorkor and acrodermatitis enteropathica is rarely reported. We herein reported a case of a 2-year-8-month-old boy with Hirschsprung’s disease who had the classic signs of both kwashiorkor and acrodermatitis enteropathica. Administration of intravenous human albumin with oral zinc gluconate 5 mg daily resulted in complete resolution of the dermatitis within ten days.

CASE REPORT

A 2-year-8-month-old boy, full-term with adequate birth weight, presented with a 2-day history of generalized desquamations over his trunk and limbs. He was diagnosed with Hirschsprung’s disease since birth and had received total colostomy and Duhamel’s pull through with anastomosis at the age of 4 months. Thereafter, he suffered from multiple episodes of acute enterocolitis due to impaired gastrointestinal function. Growth delay and mental retardation were noted, along with a body weight of 4.7 kg and a height of 70 cm (both less than the 3rd percentile of the median, WHO reference).

Two weeks before presentation, he underwent post-prandial vomiting and watery diarrhea for 3 days. Abnormal laboratory data included elevated total white blood count (11,500/μl) with a differential of polymorphs 38%, lymphocytes 55% and monocytes 8%, increased C-reactive protein level (1.7 mg/dl, normal < 1.0 mg/dl), normocytic anemia (hemoglobin 10.2 g/dl; MCV 85.2 fl), and hyponatremia (129 mmol/L, normal: 135-147 mmol/L). Under the impression of enterocolitis associated with the underlying Hirschsprung’s disease, he was admitted to receive systemic antibiotic therapy and electrolyte correction. However, 10 days after admission, generalized skin desquamation appeared. Despite the application of petrolatum, the skin lesion deteriorated. Generalized diffuse brownish peeling scales over the trunk and four limbs (Fig. 1A) with eczematous crusts on the face, predominantly in the perioral area and perianogenital region were noted (Fig. 1B & 1C). Thin, fragile, and light-colored hair was also found. Preceding the brownish peeling scales were erythematous plaques noted on his back. Edema of the extremities was observed. A skin biopsy taken from the junction of erythematous and pigment-
ed plaques on his back demonstrated parakeratosis, hyperkeratosis, irregular acanthosis, and mild spongiosis in the epidermis. Mild perivascular mononuclear infiltrate was demonstrated in the superficial dermis (Fig. 2). The serum albumin was 1.7 g/dL (normal: 3.7-5.3 g/dL). Zinc deficiency was confirmed with a low serum zinc level of 45 ug/dl (normal 70-140 ug/dl). The blood levels of vitamin B12 and folate were within normal range. Bacterial cultures of blood, urine and stool were negative.

We prescribed oral zinc gluconate supplements (5 mg daily) and intravenous human albumin (1 g/kg for three days). Clinical improvement occurred within 3 days, and complete skin healing was achieved within 10 days (Fig. 3). He remained on oral zinc supplements for 3 months, and there was no subsequent skin eruption in the following two years. However, enterocolitis associated with the poor gastrointestinal function of Hirschsprung’s disease recurred four times till now.

DISCUSSION

Kwashiorkor, a nutritional disease of children relative to maize diet, was first described by William in 1935. The term “kwashiorkor”, derived from the Kwa language of Accra, Ghana, indicates the disease of a weaned baby when the next one is born. Kwashiorkor is now thought to be severe protein-energy malnutrition in which the caloric intake remains adequate, whereas protein amount is deficient and carbohydrates are relatively excessive. It is more prevalent between 6 months and 8 years of age in developing countries, especially in the areas where the diet mainly consists of corn, rice, or beans. Symptoms start with a gradual failure to thrive, irritability, photophobia, muscle wasting, diarrhea, lack of skeletal and mental development, edema of the extremities, and skin lesions. The diagnosis is based on a characteristic skin rash and clinical presentations. Skin findings comprise hypo- or hyperpigmented patches and erosions, with a predilection in the pressure zones and extremities. The surfaces of the patches desquamate and delineate a characteristic “flaky paint” dermatosis. Hair becomes dry, sparse, with color changes including a diffuse whitening or reddish hue, which are particularly representative. Some patients’ hair have “flag sign” paralleled to bands of dark and light colors that demonstrate the periods of adequacy and worsening in their nutrition condition. The deficiency of protein absorption leads to edema of the extremities and bacterial or parasitic infection. The findings of edema, hypoalbuminemia, and characteristic dermatitis distinguish it from starvation or marasmus, resulting from total energy insufficiency.

Kwashiorkor is associated with extreme poverty in developing countries. It is also reported to occur in other chronic malabsorptive or malnutritional conditions such as anorexia nervosa, cystic fibrosis, partial gastrectomy, food aversion, acquired immunodeficiency syndrome, and malignant histiocytosis.

The exact mechanisms of kwashiorkor are unclear. Recent biochemical researches in kwashiorkor include increased oxidative and nitrosative stress, elevated prostaglandin E2, and dietary toxins like aflatoxins in starchy foods. Some hypotheses consider it a multiple deficiency syndrome related to a lack of essential amino acids, vitamins, and trace elements, especially zinc.

Liu et al. analyzed 12 cases related to chronic illness or nutritional ignorance in America. Five of 12 patients had a low zinc lev-
el, and 3 of the 5 cases improved without zinc supplement. Zinc deficiency has been thought to be a contributing factor for kwashiorkor, while protein malnutrition is the primary reason for it. The role of zinc deficiency was highlighted by Golden et al. Seven of 8 patients with kwashiorkor were reported as showing more rapid healing on the topical zinc-supplemented side of the lesion than the placebo-supplemented side. He suggested that the skin lesions of kwashiorkor and acrodermatitis enteropathica both seem to be caused by a deficiency of zinc.

Acrodermatitis enteropathica is a rare disease caused by zinc deficiency, either hereditary by autosomal recessive transmission or acquired. Acquired acrodermatitis enteropathica usually develops secondary to total parenteral nutrition. The typical cutaneous manifestations include acral dermatitic, eczematous, desquamative, and erosive lesions around the body orifices, such as perioral and anogenital regions, alopecia, and diarrhea. The diagnosis is based on clinical features and should be confirmed by lower plasmatic zinc concentrations. In the literature review, only one case report reveals a 26-year-old woman with anorexia nervosa exhibiting clinical signs of both kwashiorkor and acrodermatitis enteropathica. Hypoproteinemia and hypozincemia were both noted in the patient, and zinc replacement together with intravenous human albumin resulted in complete remission of the skin lesion in 2 weeks.

Hirschsprung’s disease, also known as congenital megacolon, is a motor disorder of the colon, which results from the failure of neural crest cells migration during colonic development (during the fifth to twelfth weeks of gestation). Consequently, the affected segment of the colon fails to relax, causing functional obstruction. The disorder occurs in 1: 5000 live births with a male predominance of 3.8 to 1.0. Seventy-five percent of cases are limited to the rectum and sigmoid, and only 8% of cases have total colon involvement. The mainstay of treatment is surgery to excise the affected colon and to bring the normal ganglionic bowel down to the anus and preserve sphincter function. However, recurrent enterocolitis owing to intestinal stasis is a major problem postoperatively. To our knowledge, the coexistence of kwashiorkor and Hirschsprung’s disease has never been presented before. Our patient had Hirschsprung’s lesion involving the entire colon. This severe form of Hirschsprung’s disease might have aggravated the intestinal function, leading to the severe dermatitis. For the reason, strict monitor of the growth status and adequate oral feeding with nutritional supply are necessary for those patients in postoperative courses.

Our patient presented with “flaky paint” dermatitis and hypopigmented, spare, fragile hair, which are pathognomonic of kwashiorkor. The course of disease was acute. He did not have a similar skin problem even though the albumin levels were as low as 2.8 g/dL one year before kwashiorkor attacked. However, during the active stage of kwashiorkor, he had the lowest level of albumin of 1.7 g/dL. Sudden breakdown of protein may bring about the disease. Dramatic remission of skin lesions after zinc supplement suggesting zinc deficiency might be an inducing factor for kwashiorkor, since hypoalbuminemia has existed for a period of time.

In conclusion, kwashiorkor is a worldwide disease not only in developing countries, but also in developed ones. The diagnosis depends on the clinical presentations. Some patients with kwashiorkor have low plasma zinc, however, further studies are required to answer the role of zinc deficiency in kwashiorkor, since zinc replacement contributes to skin recovery. Recognizing the features of kwashiorkor can enable dermatologists to treat patients timely and avoid detrimental consequences.

REFERENCES