

at least for the present, that the survey results indicate that 47,XYY subjects have no port-wine stain, or that the authors did not care about it.

Further dermatological studies on 47,XYY males and further chromosome studies on patients with port-wine stains are clearly needed.

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Kwashiorkor-like Zinc Deficiency Syndrome in Anorexia Nervosa¹

Serban A. Esca, Wilhelm Brenner, K. Mach, and Fritz Gschnait

Department of Dermatology I, University of Vienna, Vienna, Austria

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Abstract. This report deals with a 26-year-old white woman exhibiting signs of both Kwashiorkor (marasmus, pallor, hypopigmentation of hair and hepatomegaly) and acrodermatitis enteropathica (eczematous dermatitis predominantly on acral areas). Clinical and laboratory examinations excluded malabsorption syndrome and glucagonoma syndrome and revealed hypoproteinemia and marked zinc deficiency. Psychiatric examination disclosed anorexia nervosa. Substitution therapy led to rapid clearing of the skin lesions.

Key words: Anorexia nervosa; Zinc deficiency; Kwashiorkor; Acrodermatitis

Dermatological symptoms following zinc deficiency have been reported to occur in several conditions, including acrodermatitis enteropathica (1), malabsorption syndrome (11, 15), chronic alcohol abuse (15) and during long-term total parenteral nutrition (2,3). The present paper deals with both Kwashiorkor-like (4) and acrodermatitis enteropathica-like (5) skin changes in a patient with anorexia nervosa.

CASE REPORT

A 26-year-old female teacher was admitted to hospital because of dermatitis, diffuse alopecia and weight loss. She was well until 4 months prior to admission, when she stopped normal food intake and started to drink abnormal amounts of alcohol, her meals being confined to pickled cucumbers and wine only. Over the following 4 months she lost 20 kg of body weight and developed a skin rash. In addition, diffuse hair loss occurred 4 weeks prior to admission.

On physical examination she appeared malnourished and pale. An eczematoid dermatitis involved the hands, the lower back, buttocks, perianal and perigenital areas (Fig. 1a) and the lower legs (Fig. 1c), where erythematous, moist, erosive, sharply but irregularly outlined patches with marginal scaling were prominent (Fig. 1a).

The fingers showed a thin erythematous slightly scaly atrophic skin. The scalp hair was loose, thin and exhibited a conspicuous light discoloration (Fig. 1b) of the lower portion of the hair shaft. A similar pattern of hypopigmentation (Fig. 1b, inset) was found on the eyebrows and eyelashes. Except for pitting oedema of the ankles and lower legs there were no other pertinent skin changes. On palpation, her liver was found to be increased in size.

On routine laboratory examination there was hypokalemia (2.8 mval/l) hypocalcemia (7.7 mg/100 ml) hypoproteinemia (5.3 g/100 ml) and a pathologic liver profile: total bilirubin 2.2 mg/100 ml, serum alkaline phosphatase 196 U/l, SGOT 65 U/l, and SGPT 30 U/l.

Other laboratory data including CBC, urinalysis, fasting blood sugar, oral glucose tolerance test, serum iron, iron binding capacity, blood urea nitrogen, serum creatinine, serum protein electrophoresis, immunoelectrophoresis, plasma amino acids, creatinine clearance were within normal limits.

Schilling-test, Gordon-test and xylose-test did not reveal any signs of malabsorption. Plasma glucagon levels as measured repeatedly by radioimmunoassay were within normal ranges.

Serum zinc levels determined repeatedly by atomic absorption spectrophotometry were below 40 µg/100 ml (normal 90 ± 20 µg/100 ml). Potassium hydroxide preparations and culture from eroded areas did not show fungi. On histologic examination of the affected skin there was psoriasiform dermatitis with hyperkeratosis, parakeratosis, spongiosis and polymorphonuclear and round cell inflammatory infiltrate in the upper dermis (Fig. 2). Direct and indirect immunofluorescence examinations using anti IgA, IgG, IgM, IgE and C₃ antisera were negative.

EKG, chest X-ray, GI series, IVP, coeliacography, ultrasound examination of pancreas, kidney scan and laparoscopy proved normal. Liver biopsy revealed mild fatty degeneration but no gross pathology. Psychiatric

¹Dedicated to Prof. Dr Anton Luger on the occasion of his 60th birthday.

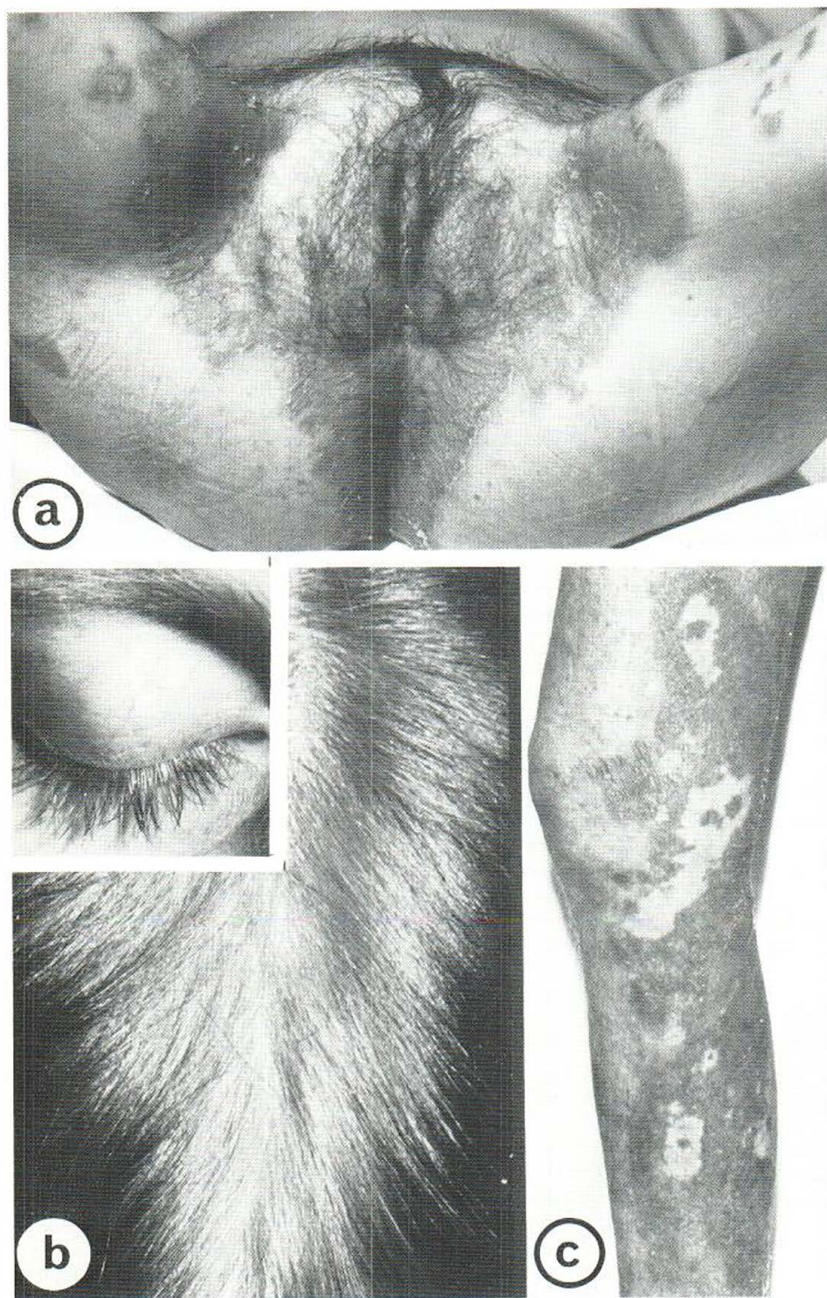


Fig. 1. (a) Perigenital region showing sharply outlined erythematous patches with marginal scaling. (b) The hair of the scalp exhibits light discoloration of the lower portion of the hair shaft. Inset: a similar pattern of hypopigmentation on the eyelashes. (c) Erythematous, sharply but irregularly outlined patches with marginal scaling on the legs.

examinations disclosed the nervous cause of anorexia which was due to the patient's difficulties in the family.

Zinc replacement therapy with Solvezink 45 mg tablets ($ZnSo_4$) three times a day together with i.v. administration of human albumin resulted in prompt healing of the skin lesions after a few days' therapy and, after 2 weeks, complete clinical remission occurred. For topical treatment, emollients only were used, which did not influence the

skin condition prior to zinc administration. Laboratory data, which were pathologic prior to treatment, returned to normal.

After discharge from hospital the patient was left on three tablets Solvezink per day for a further 4-week period during which psychiatric treatment was started, resulting in normal life activities including normal food intake. The patient regained 20 kg of body weight, the

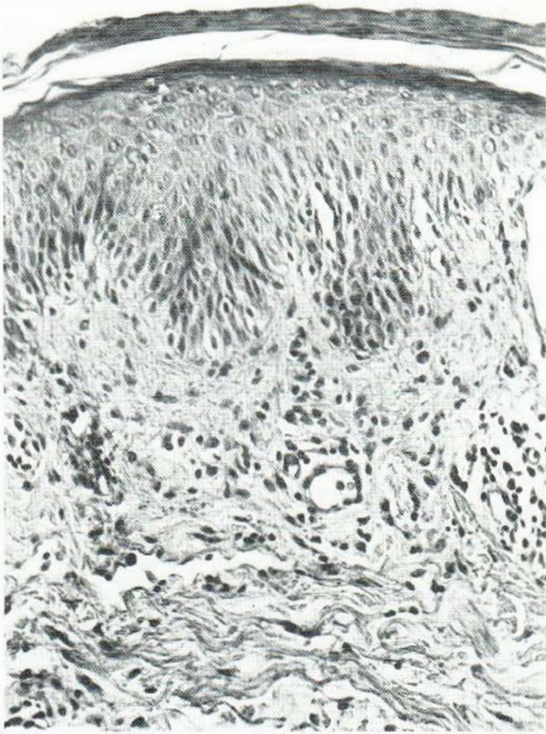


Fig. 2. Biopsy from the affected perigenital region: Psoriasiform dermatitis with parakeratosis, spongiosis and polymorphonuclear and round cell inflammatory infiltrate in the upper and mid-dermis. (H & E, $\times 125$).

pitting oedema of the lower legs disappeared and normal pigmented hair was now growing. Up to 6 months after hospitalization, serum zinc level and skin condition remained normal.

DISCUSSION

Zinc deficiency states were first recognized and related to skin disorders in animals in 1955 (13). In 1961 Prasad et al. described iron deficiency anemia, hepatosplenomegaly, hypogonadism, dwarfism and geophagia due to zinc deficiency in young Iranians and Egyptians whose food intake contained large amounts of phytates—substances later recognized as producing hypozincemia by chelation of zinc (6).

In 1973 Barnes and Moynahan discovered zinc deficiency to be the ultimate cause of acrodermatitis enteropathica and since that time research in endogenous zinc metabolism has led to a better understanding and earlier recognition of zinc deficiency causes and clinical manifestations.

Zinc was found to play a major role in fundamen-

tal enzymatic processes throughout the body, such as DNA, RNA and collagen synthesis (9). There are now over 70 enzymes known to be either zinc metalloenzymes or to require zinc as co-factor (9).

Acquired zinc deficiency has been mentioned (6) to occur possibly in a variety of conditions, such as renal diseases with protein-losing nephropathy, hemolytic anemia, catabolic conditions with muscle wasting, alcoholic cirrhosis, etc. Dermatologic changes due to hypozincemia have been reported in patients on long-term parenteral nutrition (2,3), in chronic alcohol abuse (15, 16) and in malabsorption syndromes (11, 16). Acquired zinc deficiency was also studied experimentally in volunteers on a restricted zinc intake (8) leading to a decrease in zinc concentration in plasma, erythrocytes, leukocytes and urine. It produces early signs such as anorexia, hypogeusia, hyposmia and, if unrecognized, may lead rather late to the development of skin lesions as seen in acrodermatitis enteropathica.

In the present case hypoalbuminemia due to anorexia nervosa resulted in hypoproteinemia and hypozincemia and this explains the development of a condition including the symptoms of both Kwashiorkor—such as marasmus, pallor, depigmentation of hairs, hepatomegaly—and of acrodermatitis enteropathica.

Although the mechanism of zinc absorption and zinc metabolism is not yet completely understood, it seems that ingested zinc after resorption is bound primarily to albumin. The replacement of both deficient substances (albumin and zinc) led to normal zinc bioavailability and thus resulted in rapid clearing of the skin lesions.

In the present case the main differential diagnosis included necrolytic migratory erythema the dermatological manifestation of the glucagonoma syndrome (12) which was disproved by normal glucose metabolism, normal glucagon levels in the serum and the failure to detect a pancreatic-tumour by ultrasound examination, celiacography and laparoscopy.

The question remains to be answered whether the skin changes in true african Kwashiorkor are due solely to hypoproteinemia, or also to hypozincemia.

ACKNOWLEDGEMENT

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Inflammatory Linear Verrucous Epidermal Nevus (ILVEN)

Inger Skoven

Department of Dermatology, Odense University Hospital, Odense, Denmark

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Abstract. A 12-year-old boy with inflammatory linear verrucous nevus is described. The lesions were pruritic and consisted of small, erythematous, slightly scaling papules coalescing to form linear, lichenified and excoriated plaques. The histological picture was psoriasiform but features of eczema were also seen.

Key words: Linear epidermal nevus; Inflammatory psoriasiform features

In 1971 Altman & Mehregan, basing their investigation on 25 cases, described inflammatory linear verrucous epidermal nevus (ILVEN) as a distinct clinical and histological variety of linear verrucous nevus or nevus unius lateris (1). Sporadic cases had earlier been reported as "cases for diagnosis" (4), atypical cases of linear psoriasis (2, 6), or as psoriasiform linear nevus (7). Additional cases have been presented in a few reports (3, 5, 8) following establishment of the condition as a distinct disease entity.

The following features are characteristic of ILVEN:

1. Unilateral, usually pruritic lesions, consisting of small, discrete, erythematous, slightly verrucous, scaling papules which tend to coalesce to form linear plaques.
2. Clinical and histological resemblance to either psoriasis or eczema.
3. Absence of typical psoriasis.
4. Female predominance of 4:1.
5. Early onset of lesions, 50% before the age of 6 months, 75% prior to the age of 5 years.
6. Minimal response to treatment.

CASE REPORT

A 12-year-old boy was admitted to the Department of Dermatology, Odense University Hospital, with an extremely pruritic, linear lesion on his right leg extending from the ankle to the loin (Fig. 1). There was no family history of psoriasis or atopic disease. The lesion had developed at the age of 6 years, starting in the popliteal area