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Dermatological Study of 47,XYMales

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Abstract. Dermatological features of five 47,XYMales are presented. Port-wine stains were observed in 2 cases. As the incidence of port-wine stains among the general population is believed to be 0.5%, this result would seem to indicate that the association of 47,XYMales with port-wine stains is more than a coincidence, though the survey of previous studies failed to reveal any 47,XYMales cases with port-wine stains.

A male with 47,XYM karyotype was first described by Sandberg et al. (2). Though more than 370 cases have been reported since then, no characteristic features of those males have been detected, except

Table 1. *Dermatological features of 47,XYMales in the literature*

Numbers indicate cases with positive findings

Acne	26
Varicose vein/ulceration	14
Dry skin	2
Hyperhidrosis	2
Wrinkly face	2
Baldness	2
Other (pigmented skin, neurodermatitis, telangiectasia, café-au-lait patch, tinea pedis, sebaceous adenomata, white macula, psoriasis, nevus)	

for high stature. Little is known about the dermatological features in 47,XYMales, the available data being limited to isolated case reports or reports involving only a few subjects—apart from the study of Voorhees et al. (3), who found an unusually early onset of acne (at 3 years of age) in 47,XYMales. They later (4) suggested that 47,XYMales often have nodulocystic acne, an observation based on a survey of patients with nodulocystic acne. They found 47,XYMales eleven times more frequently than expected.

In this study, the dermatological features of 5 47,XYM cases are described and those in a previous study are reviewed.

MATERIALS AND METHODS

The material consisted of 5 47,XYMales found among juvenile delinquents (1). The age of the subjects ranged from 16 to 19 years. The parents of Case 1 were first cousins once removed. Those of Case 3 were first cousins. The others were unrelated. All cases were examined at dermatological interview and by inspection by one of the authors (H. N.).

In addition, dermatological features of 47,XYM subjects in previous reports were surveyed based on the compilation data in our laboratory (data available on request).

RESULTS

Three patients (Cases 3, 4, 5) displayed a mild form of acne vulgaris which emerged in adolescence. No patient showed nodulocystic acne. Port-wine stains were observed in 2 cases (Cases 3, 4). The port-wine stain of Case 3 was on the upper abdomen, 4.0×7.0 cm in size (Fig. 1). The port-wine stains of case 4 consisted of a relatively large patch (2.0×1.5 cm) and some small macules (0.5×0.5 cm) on the right forearm (Fig. 2). Another case (Case 5)



Fig. 1. Port-wine stain of Case 3.



Fig. 2. Port-wine stains of Case 4.

had a history of atopic dermatitis between the ages of 2 and 6 years.

Table I shows the dermatological features of 47,XYY subjects in the previous reports. Of 375 cases, only 50 were described as to dermatological features.

DISCUSSION

Acne vulgaris was a common finding in this study and in previous studies. However, this was also common in the general population, especially during adolescence. Thus this finding might not indicate anything of particular significance.

On the other hand, port-wine stains were observed in 2 cases out of 5. As the incidence of port-wine stains among Japanese general population is believed to be 0.5% (Hidano, F., personal communication), this result would seem to indicate that the association of 47,XYY subjects with port-wine stains is more than mere coincidence. One problem is that no 47,XYY subjects with port-wine stains have been reported in the previous studies. We assume that this is the reflection of authors' indifference to dermatological features, since in only 50 out of 375 cases were dermatological features described. Anyway it is difficult to maintain,

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

Further dermatological studies on 47,XXX 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

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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.