LETTERS TO THE EDITOR

Lichen Ruber Planus Following the Administration of Human Anti-hepatitis B Virus Immunoglobulins

Sir,
A 50-year-old nurse was referred to our department because of the onset, about 3 weeks previously, of many small, flat-topped, polygonal, pink, itching papules, many of them forming plaques. The smooth surface of the lesions presented a network of white lines. The wrists, lower back and ankles were mainly affected, but the whole skin was involved too. There were many white streaks on the mucous membranes of the cheeks. The clinical picture was the peculiar one of lichen ruber planus (LRP). One month before the onset of the skin lesions the patient, working at the internal medicine department, had been given anti-hepatitis B virus (HBV) human immunoglobulins after she had accidentally got stung by a used needle.

Upon anamnesis she denied any cutaneous or internal disease. Annual serologic tests for HBV and HCV were always negative and she had never received HBV vaccine. Medical examination was negative for other diseases. Routine laboratory investigations, including liver enzymes and bilirubin, were all normal. She had only a low titre of anti-HBs antibodies (37 mIU/ml) as the result of the previously administered immunoglobulins. The other HBV markers, anti-HCV antibodies, organ and non-organ-specific autoantibodies were negative. The patient did not consent to a skin biopsy. She was not taking any drugs and had not been exposed to known LRP-inducing chemical substances (1). The relationship between chronic HBV and HCV and LRP is reported in the literature (2, 3), as are some rare cases of LRP induced by plasma-derived and recombinant HBV vaccines (4, 5). We did not find any cases of LRP following the administration of anti-HBV human immunoglobulins. Our case may support Rebora’s hypothesis (5) about the sharing of common epitopes between HBV and the keratinocytes. An immune response against those antigens would clinically develop as LRP.

REFERENCES

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Treatment of Lipoid Proteinosis with Eretinate

Sir,
Lipoid proteinosis is a rare nosologic entity, also called Urbach–Wiethe disease or hyalosis cutis et mucosae. The disease is characterised by the deposit of hyaline-like materials in the skin, mucous membranes and other tissues. The etiopathogenesis of lipoid proteinosis has not yet been clearly established, but it is regarded as an autosomal recessive genodermatosis, with variable expression. Parental consanguinity has been described rather frequently (1). The first manifestation of the disease is hoarseness present soon after birth or early in infancy, caused by intratracheal deposits of hyaline material in the vocal cords. Skin lesions such as papules, nodules and plaques appear shortly afterwards, and with time calcifications appear in the skin. Typical papules are present on the upper and lower eyelids in about 50% of the cases (2). We here report a good effect of etretinate on the skin lesions.

CASE REPORTS

Case 1
A 36-year-old woman had been suffering from hoarseness and dysphonia since birth, and from her first year of life she had had papules, vesicles and later scars. There was no family history of skin diseases but consanguinity was present. She is rather short of stature; her face is hypomimic and coarse, with scars on the forehead, cheeks and sides of the head. The eyelids presented along the margins typical white papules. The joints and the knees there were verrucous plaques. There were crustated plaques and scars on the trunk (Fig. 1). The hair was sparse. The tongue was thickened and affixed to the floor of the mouth.

Indirect laryngoscopy revealed thickened and uneven vocal cords. X-ray showed intracranial calcifications overlying the sella turcica. A biopsy of the skin showed the characteristic features of lipoid proteinosis with orthohyperkeratosis and hyaline deposits in the dermis, which gave a strongly positive reaction with PAS.

Investigations showed normal blood count, serum protein, blood urea, liver function tests and serum lipids.

Previous therapy with dermabrasion had produced little and only temporary benefit. After a formal verbal consent had been obtained from the patient, she was treated with etretinate 1.0 mg/kg daily for 2 months and then with 0.75 mg/kg. At the follow-up after 4 months the examination revealed a substantial improvement of the verrucous lesions on the knees and elbows and of the papules on the palms. She felt the skin to be more elastic. As the plasma level of cholesterol and triglycerides increased (6.8 mmol/l and 2.4 mmol/l, respectively) and also the transaminases (ALT 31 µ/l, AST 28 µ/l), the therapy was stopped. After 2 months the lesions relapsed.
Case 2

A female aged 40 years had had skin lesions present from early infancy and hoarseness from birth. The diagnosis of lipoid proteinosis was confirmed by histology. The family history revealed parental consanguinity, and some members of the family had epilepsy. As a child the patient was diagnosed as having epilepsy, and she had been treated with phenitooin ever since then.

The skin was pale and on the margins of the eyelids were present typical waxy papules. The tongue was pale, depapillated, hard and thickened; along the lips there were whitish papules and fissures (Fig. 2). Indirect laryngoscopy showed irregular, thickened vocal cords and X-ray showed calcifications overlying the sella turcica, and in both maxillary sinuses. Blood count and all biochemical analyses were normal.

The patient was treated with etretinate 1.0 mg/kg daily, for 2 months and then with 0.5 mg/kg for 4 months. The lesions on the elbows, knees and on the hands improved. She believed that her skin looked better and became more elastic. Her skin and her lips became dry, which was diminished by emollients.

Although these preliminary observations showed an improvement of the disease, one might not expect a complete regression of it. Nonetheless our trial clearly demonstrated that retinoids may act on the cutaneous lesions in lipoid proteinosis. Indeed, the observation that the skin manifestation responds to such a brief treatment period is encouraging, and perhaps with a longer course of therapy, a better improvement would be seen.

REFERENCES


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