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Case Report

Hidroa vacciniforme

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Abstract

Hidroa vacciniforme is a rare chronic photodermatosis with unknown cause in which lesions appear with exposure to sun. Early diagnosis and treatment of this disease which is characterized with recurrent vesicles and bullae on areas which are exposed to sun including the face and distal parts of the extremities is important, since it recovers by leaving scar. Here, a six-year old girl who presented to our clinic with eruptions on the face for three years and who was diagnosed with hidroa vacciniforme was presented in accompaniment of current information because of the rarity of the disease.

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Key words: Childhood, photosensitvity, hidroa vacciniforme, histopathology

Introduction

The differential diagnosis of skin rash occuring with exposure to sunlight in the childhood may be difficult. These lesions may be triggered by exposure to drugs or chemicals or they may be a part of a clinical picture belonging to a systemic or immune disorder. In addition to these diseases, idiopathic photodermatoses including solar uritcaria, polymorphous light eruption and hidroa vacciniforme should also be considered in the differential diagnosis. Although idiopathic photodermatoses are a considerably wide group, the diagnosis can be made in the childhood by taking a careful history, clinical examination and phototests when necessary (1). Here a patient with hydroa vacciniforme (HV) which is one of the photodermatoses and for which early diagnosis and treatment is important, since it improves by leaving scar was presented with the consent of the family.

Case

A 6-year old female patient presented with rash which especially occured with exposure to sunlight on the back of the hand, forehead and face and which healed by leaving scar for the last three years. In her history, it was learned that her complaints started in the spring three years ago and lasted throughout the summer. The lesions occured as mild pruritus, erythema and vesicles in hours following exposure to sunlight, crusted in a few days and healed by leaving scar. She had no history of any morbidity or use of any photosentisizing substance. It was reported that the parents had no consanguinity or morbidity. On physical examination, papulovesicles which were necrotic in the middle were observed especially on the frontal and zygomatic regions, bullea with a diameter of 0.3 cm were observed on the auricle and 2.5 mm atrophic scars were observed on the face (Figure 1). Chain-formed cervical lympadenopathies with a diameter of 0.5 cm were found bilaterally. Other physical examination findings were found to be normal. Hemogram, peripheral smear, transaminases and 24-hour-urine porphyrine values were found to be normal. Serological examination revealed that Epstein Barr Virus (EBV) viral capsid antigene IgM, EBV viral capsid antigene IgG and EBV nuclear antigene IgG were negative. On histopathological examination of skin biopsy,

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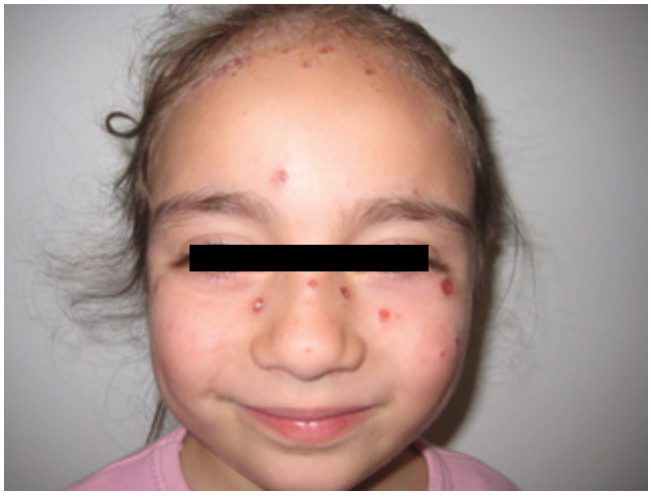

Figure 1. Papulovesicles which are necrotic in the middle in the frontal and zygomatic regions, bullae with a diameter of 0.3 cm on the auricle and 2-5 mm atrophic scars on the face

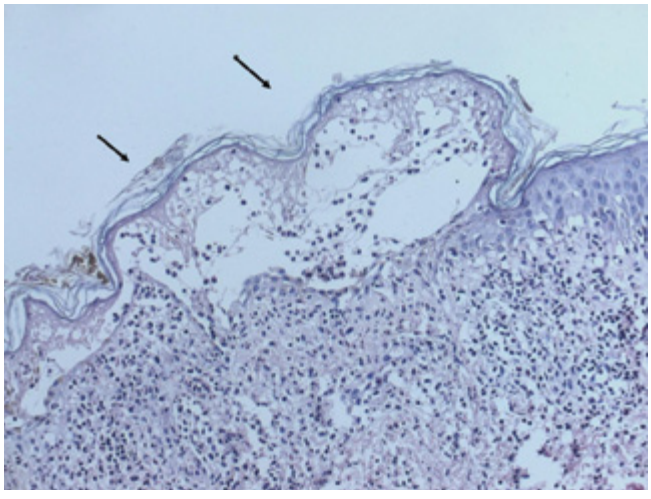

Figure 2. Pustule formation in the epidermis and necrosis in the keratinocytes which constitute the pustule roof (arrows). Intensive mixed type inflammation in the dermis (H-E, x200)

subcorneal pustule formation was observed in the epidermis and sponge-like appearance, lymphocyte and neutrophil exocytosis was found in the areas adjacent to the pustular lesions. Edema in the dermis which appeared as a thin line included the periperal areas of the vessels and intensive inflammatory cell involvement (neutrophils, leukocytes and lymphocytes) was observed in the interstitium (Figure 2). Lymphoid cells were shown to be polyclonal immunohistochemically by applying CD3 and CD20. A diagnosis of HV was made in the light of history, typical examination and laboratory findings in the patient who had no evidence of neoplasm. Mechanical protection from sunlight, wide-spectrum suntan creams and syrup containing fish oil were recommended to the patient. In the follow-up, no new lesion was observed on the areas which could be protected from sunlight.

Discussion

Hydroa vacciniforme is a rare chronic photodermatosis with unknown cause in which lesions appear with exposure to sun. The disease which was described by Bazin in 1862 for the first time is characterized with bullae and vesicles recurring on the areas which are exposed to sun and its typical characteristic is to leave scars similar to varicella. The word hydroa means water in greek and reflects the vesiculo-bulleous nature of the lesion. The word vacciniforme is derived from the word "vaccinum" which means vaccine in Latin expresses healing by leaving a scar like the scar of a vaccine (1, 2).

The incidence of hydroa vacciniforme is approximately 0.34/100 000 and it has been reported equaly in both genders. However, it has been reported that it has a later onset in men and the disease lasts longer in men compared to women (3). Our patient was compatible with the literature in terms of age of onset and gender. The etiology of hydroa vacciniforme is not known and its association with human leukocyte antigenes DRB1 locus has been demonstrated (4). Hypersensitivity to 320-390 nm ultraviolet A beams may be involved in the pathogenesis (5). In addition, the relation of HV with EBV has been emphasized in recent years (4, 6). Iwatsuki et al. (6) showed the presence of small nuclear ribonucleic acid (EBER) cells which are coded by EBV in skin involvement in 6 patients with clinical and histological typical HV. In addition, they proposed that typical and atypical HV were the forms included in the same disease spectrum as EBV-related lymphoproliferative diseases. Similarly, in a study they conducted in 2006, they showed that the amount of EBV-deoxyribonucleic acid in mononuclear cells in the peripheral blood was increased, though no hematologic disorder was present in patients with typical HV and HV-like lesions (7). On the other hand, an increase in EBV-deoxyribonucleic acid levels together with marked "natural killer" cell lymphocytosis and an increase in problems including hypersensitivity to mosquito bites, hemophagocytic syndrome and chronic active EBV infection have been found in the severe HV group (7).

However, patients with EBV-related HV-like lesions also have atypical skin lesions on the areas which are not exposed to sun and accompanying systemic findings and these patients have been generally reported from Asia. In such patients, there is a possibility of development of hematological malignencies. Therefore, it has been accepted that such a severe clinical picture with atypical localization and systemic findings should be evaluated as a picture which is different from typical HV, though it has clinical and histopathological findings similar to HV (8).

Clinically, it is characterized with tense edematous papules and vesicles. The lesions are necrotized in the middle in time. They crust and heal by leaving scar. Ocular involvement has been reported very rarely and it may be observed as conjunc-

tivitis, vesiculer lesions in the conjuntiva, corneal involvement and keratouveitis (9). Rarely, photo-onycholisis in the nails may also be observed. Hann et al. (10) reported a case of HV wich leaved contractures leading to severe scarring on the face and deformation in the fingers. Although the disease heals spontaneously in the adulthood, the fact that the scars are permanent creates an important problem. Kim et al. (11) reported a patient with HV who was diagnosed at the age of 22 years with serious disorder as severe as mutilation and severe scars in the auricle. Our patient was diagnosed early and had no nail or ocular involvement. She had few scars and no systemic problem accompanied. Therefore, the prognosis can be expected to be good with an efficient protective treatment.

In the differential diagnosis, polymorphous light eruption, bulleous lupus erythematosus and erythropoetic protoporphyria should be considered as well as bulleous impetigo and herpes virus infections. However, the differential diagnosis can be easly made with the history, age of onset, physical examination findings, old scars and normal laboratory tests (2). Although careful history taking and physical examination findings are usually sufficient for the diagnosis, histopathology or test with ultraviolet may be necessary. Histopathologically, intradermal reticular degeneration and cell necrosis are observed. A T-cell lymphoma type which causes to necrotic lesions on the face should also be considered in the differential diagnosis. In this lesion, presence of intensive atypical lymphoid cells is observed histopathologically. Neoplastic cells are stained CD3 (+), CD20 (-) (12). In our patient, no such increase in atypical cells was observed and lymphocytic cells were positive both with CD3 and CD20. Stimulation test with ultraviolet A is important especially in the differential diagnosis of adult patients (3, 13). However, the family did not accept phototest because of its tendency to leave scar. Our patient was diagnosed with HV with histopathologic examination and clinical findings.

In treatment of hydroa vacciniforme, protection from sun is essential. In addition to_mechanical protection from sun, mineral suntan creams are recommended (14). In a study performed in patients with hydroa vacciniforme, it was shown that the disease decreased the quality of life in children because it prevented playing outside (15). In patients in whom no sufficient response can be obtained, phototherapy (ultraviolet B TL-01), beta carotene, diet rich in unsaturated fatty acids, cyclosporine A and antimalarial drugs may be used (3, 16, 17). Lysell et al. (18) reported a decrease in the frequency and severity of attacks in three of four patients they treated with acyclovir/valacyclovir.

Protection from sunlight with clothes, wide-spectrum suntan creams and syrup containing fish oil were recommended to our patient. Development of new lesions decreased with treatment compliance of the patient and the family. Our patient was a typical HV case with both clinical and laboratory findings and her response to treatment. In many patients, spontaneous healing is observed at the end of adolescence. Therefore, it was planned to follow up the patient with three-month intervals. However, we think that severe or treatment-resistant cases should be investigated in terms of EBV and lympoproliferative diseases and histopathological examination should also be performed.

Informed Consent: Written informed consent was obtained from the parents of the patient who participated in this study.

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