DOI: 10.2478/v10249-012-0007-6

Hydroa vacciniforme - a case report

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UDC 616.5-001.15

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Abstract

Hydroa vacciniforme is a rare, idiopathic, chronic photodermatosis that usually begins in childhood and resolves spontaneously in early adulthood. It is characterized by appearance of vesicles on sun-exposed areas. The vesicles crust and heal within one to six weeks, leaving vacciniform/varioliform scars. We report an 11-year-old boy with a 5-year history of recurrent blisters on sun-exposed areas that deteriorated each summer. He was treated with antimalarials, topical photoprotective agents, sun avoidance, dietary fish oil and supplementary doses of vitamin D3. Strict adherence to the regimen resulted in remission.

Key words

Hydroa Vacciniforme; Child; Treatment Outcome; Antimalarials; Sunlight + adverse effects; Vitamin D; Sunscreening Agents

Hydroa vacciniforme is a very rare photodermatosis of unknown etiology, with onset usually in childhood. It was first described by Bazin in 1862. The prevalence of this disorder is 0.34 per 100.000 children (1, 2).

Case report

An 11-year-old boy was admitted with a 5-year history of recurrent vesicles on photoexposed areas that worsened each summer. There was no family history of photodermatosis. He was firstly deteriorated with hydroa vacciniforme at the age of 9 years and treated with beta-carotene, omega-3 polyunsaturated fatty acids and topical photoprotective agents. The patient's history revealed that photoprotection was not adequately applied.

On examination, the boy presented with multiple hemorrhagic vesicles, erosions and crusts, as well as oval, pigmented, atrophic scars on the face (Figures 1a and 1b). Multiple crusted lesions and erosions were also observed on earlobes. Cursts, violaceous scars and residual pigmentations were present on the dorsal aspect of both hands, and there were discrete hypopigmentations on both forearms. Pediatric and ophthalmological results were normal. Abdominal echosonography was regular. Routine blood and urine laboratory tests were normal, as well as 24-hour urinary uroporphyrin level.

The treatment included synthetic antimalarials (initially hydroxychloroquine at 6 mg/kg, then chloroquine at 3 mg/kg), topical photoprotection (sunblocking creams with SPF 50+) and sun avoidance. Supplementary doses of vitamin D3 (1200 IU/day) were added. In February 2012, the patient had a relapse. Dietary fish oil was added to the regimen at pharmacological doses (4000 mg/day). Such a regimen resulted in an improvement and regression of vesicles, while the scars persisted (Figures 2a and 2b).

Discussion

Hydroa vacciniforme is a rare photodermatosis of unknown etiology that usually presents in childhood

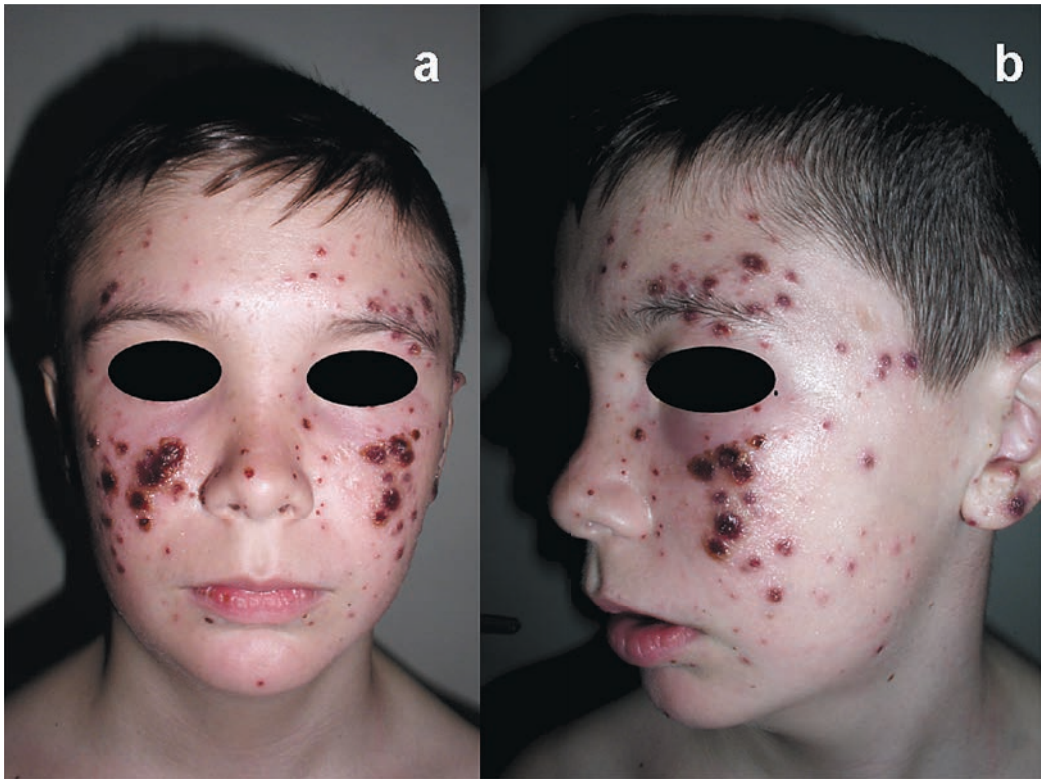

Figure 1. Hemorrhagic vesicles and crusts on the face

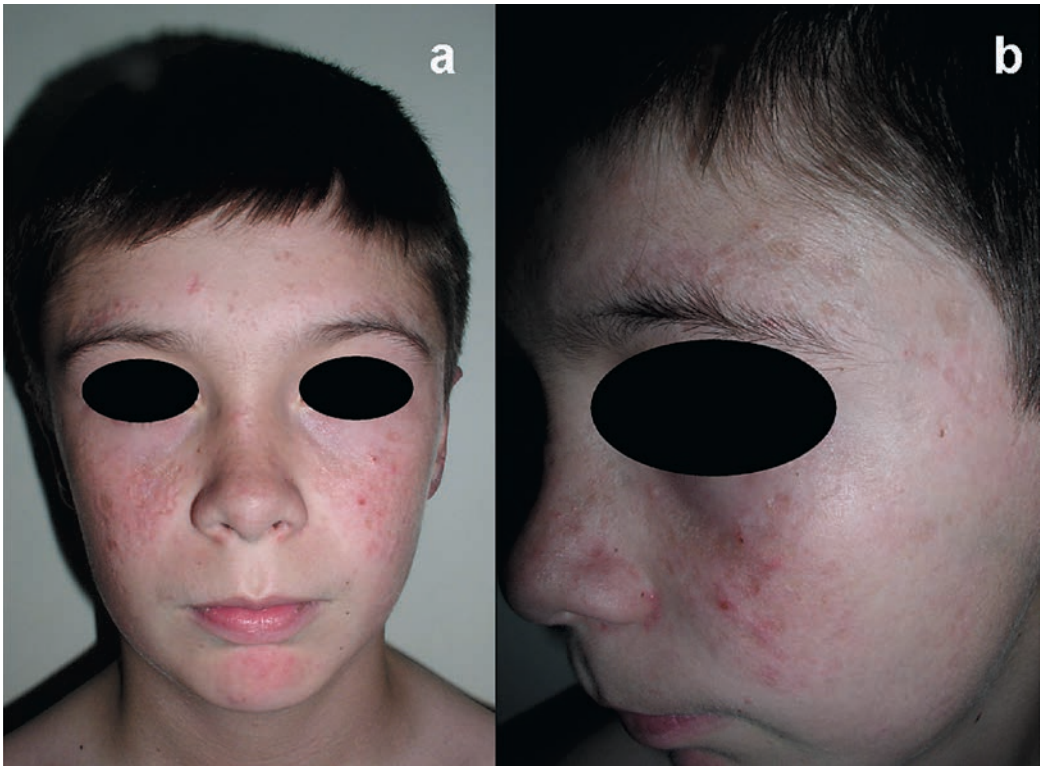

Figure 2. The patient in remission: atrophic vacciniform/varioliform scars

(3). It is characterized by itchy, stinging, erythematous rash, occurring within a few hours after sun exposure, progressing to numerous erythematous papules and plaques undergoing vesiculation (4). The vesicles tend to become umbilicated and hemorrhagic, subsequently crusted, and heal within one to six weeks, leaving depressed vacciniform scars (2). In addition, postinflammatory hypo- and hyperpigmentation may occur.

The most accepted pathogenetic hypothesis suggests ultraviolet radiation, with wavelengths between 320 and 390 nm, as a causal agent of hydroa vacciniforme, but the chromophore leading to ultra-violet-induced damage is still unknown (2). The differential diagnosis includes erythropoietic protoporphyria, congenital erythropoietic porphyria, vesicular polymorphic light eruption, actinic prurigo, and common conditions like impetigo, herpes simplex and contact dermatitis. In our patient normal 24-hour uroporphyrin level excluded erythropoietic protoporphyria and congenital erythropoietic porphyria. Polymorphic light eruption is usually non-scarring, and has a later age of onset than hydroa vacciniforme. Actinic prurigo does show scarring, but it may also involve non-sun-exposed areas, and primary lesions are papules and nodules rather than vesicles (5).

The therapy consists of UVA sunblocking agents with high SPF and protective clothing. Antimalarials, immunosuppressives, beta carotene, psoralen with UVA exposure and prophylactic UVB phototherapy have also been reported as useful in reducing outbreaks, but they are not reliable in preventing lesions. Dietary fish oil, rich in omega-3 polyunsaturated fatty acids, has also been used with success in occasional reports, with no severe side-effects reported (6).

Our patient had a good initial response to therapy, but then suffered a relapse, because photoprotection was not adequately applied. When the combined therapy was given, including antimalarials, dietary fish oil at pharmacological doses, sun-blocking creams with SPF 50+ and avoidance of the sun, the patient was introduced into remission. Supplementary doses of vitamin D3 were added because of sun avoidance.

Conclusion

Hydroa vacciniforme is a very rare disease in childhood. Timely diagnosis and introduction of adequate therapy, including antimalarials, fish oil, and strict photoprotection may enable long remission periods and normal quality of life. The disease enters stable spontaneous remission in adolescence or early adulthood.

Acknowledgement

This study was supported, in part, by the Ministry of Science of the Republic of Serbia, Grant No 175065.

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Hydroa vacciniforme - prikaz slučaja

Sažetak

Uvod: *Hydroa vacciniforme* je retka, idiopatska, hronična fotodermatoza koja obično počinje u detinjstvu i spontano regredira u ranom odraslom dobu. Prevalencija oboljenja iznosi 0.34 na 100 000 dece. Karakterišu je vezikule na zonama izloženim suncu. Vezikule se transformišu u kruste i u roku od jedne do šest nedelja postepeno zarastaju, ostavljajući vakciniformne/varioliformne ožiljke.

Prikaz slučaja: Prikazujemo dečaka uzrasta 11 godina sa promenama koje se javljaju u poslednjih 5 godina u vidu vezikula i krusti i završavaju ožiljkom, lokalizoivanim na svim fotoeksponiranim zonama. Promene su prisutne od proleća do kasne jeseni. Fizički pregled i pregled oftalmologa su bili u fiziološkim granicama.Ultrazvučni pregled abdomena, rutinske laboratorijske analize krvi i urina uključujući i nivo uroporfirina u 24-časovnom urinu su bili u fiziološkim granicama.

Dečak je lečen antimalaricima(u početku hydroxychloroquine u dnevnoj dozi od 6 mg/kg, then a potom chloroquine u dnevnoj dozi od 3 mg/kg), uz dodatak ribljeg ulja u farmakološkim dozama (4000 mg dnevno). i uz striktnu fotoprotekciju (izbegavanje sunca i kreme sa visokim zaštitnim faktorom SPF - eng. sun protection factor 50+). S obzirom da je dete sve vreme bilo zaštićeno od ultravioletnih zraka, primenjivana je i suplementacija vitaminom D3 (1200 IU/day). Striktno primenjivanje navedenih mera dovelo je do regresije promena.

Diskusija: Iako je etiologija oboljenja nepoznata, najveći etiopatogenetski značaj ima ultraljubičasto zračenje talasne dužine od 320 and 390 nm, ali je hromofora koja izaziva oštećenje još uvek nepoznata. U dierencijalnoj dijagnosi treba isključiti oboljenja kao što su: eritropoetska protoporfirija, kongenitalna eritropoetska porfirija, vezikulozna polimorfna svetlosna erupcija, aktinični prurigo, impetigo, herpes simpleks i kontaktni dermatitis. Kod našeg pacijenta isključili smo postojanje eritropoetske protoporfirije, i kongenialne eritropoetske porfirije, na osnovu urednog nivoa uoporfirina u 24-časovnom urinu. Za razliku od *Hydroa vacciniforme,* polimorfna svetlosna erucija započinje u kasnijoj životnoj dobi i nije praćena ožiljavanjem, dok se aktinički prurigo karakteriše prisustvom prvenstveno papula i nodula a ne vezikla, ožiljci se mogu javiti ali proces može zahvatiti i foto-neksponiranu kožu.

Terapija: Pored lokalne primene fotoprorotektivnih krema sa visokim SPF, nošenja zaštitnog odela, primenjuju se: antimalarici, imunosupresivi, beta karoten, psoralen u kombinaciji sa UVA zracima, profilaktička UVB fototerapija. Primenom ovih lekova može se redukovati ali ne i spečiti pojava recidiva. U dijetu treba uvesti riblje ulje bogato omega-3 višemasnim nezasićenim masnim kiselinama, što može dati pozitivan efekat bez težih neželjenih dejstava. Usled neizlaganja kože suncu,u terapiju treba kao dodatak uvesti D3 vitamin.

Zaključak: *Hydroa vacciniforme* predstavlja veoma retko oboljenje dečijeg uzrasta: pravovremeno postavljanje dijagnoze, i primena adekvatne terapije omogućuje dugotrajne remisije i kvalitetan život; bolest ulazi spontano u stabilnu remisiju u adolescentoj i ranoj odrasloj dobi.

Ključne reči

Hydroa Vacciniforme; Dete; Ishod lečenja; Antimalarici; Sunčeva svetlost + neželjena dejstva; Vitamin D; Sredstva za zaštitu od sunca