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Case report: A hepatoerythropoietic porphyria treated with hydroxychloroquine

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Introduction: Hepatoerythropoietic porphyria (HEP) is a very rare type of porphyria produced by severe deficiency of uroporphyrinogen decarboxylase (UROD) inherited in an autosomal recessive manner (biallelic mutations). Clinically it resembles congenital erythropoietic porphyria (CEP), but biochemical findings are similar to those found in porphyria cutanea tarda (PCT). These patients have photosensitivity and bullous lesions on sun-exposed areas, shortened distal phalanges, sclerodermiform skin changes (similar to CEP) and severe hypertrichosis. Photosensitivity tends to decrease with age. As in PCT, HEP presents increase of urinary excretion of uroporphyrin and heptacarboxylporphyrin with an URO/COPRO ratio $>$ 5:1 and an increase in fecal isocoproporphyrin, however it's the increase in plasma and erythrocyte protoporphyrin, which helps to differentiates it from PCT. Diagnosis may be confirmed by molecular studies of UROD mutations.

Objective: We report a patient with hepatoerythropoietic porphyria treated with low dose of hydroxychloroquine.

Case: A 20-year-old African American female presents photosensitivity since childhood, hypertrichosis, hyperpigmentation (on sun-exposed areas), blistering and erosions on the dorsal hands. She had 3 siblings with the same clinical signs and her parents were first-degree cousins. Ferritin levels were normal. Serology for hepatitis and HIV were negative. Uroporphyrin (URO) was 1798 μg/24 h (RR: $<$ 25) in the 24-hour urine sample and plasmatic porphyrins were dosed: uroporphyrin (URO) was 144 μg/L (RR: $<$ 0.2) and protoporphyrin was 9.6 μg/L (RR: 0.4-4.8). Fecal porphyrins and UROD molecular studies weren't available. Hydroxychloroquine 200 mg twice a week was introduced and as the patient showed good tolerance the dose was increased to 400mg twice a week. After one year she showed clinical improvement of hyperpigmentation, bullous lesions and of photosensitivity (URO 1081 μg/24 h urine). Her hypertrichosis was removed with waxing and yet showed no injury due to skin fragility.

Conclusion: There are many misunderstanding and misdiagnosis involving hepatoerythropoietic porphyria. Topical sunscreens alone and phlebotomy are ineffective. Most treatment information is based on other forms of porphyria and as hydroxychloroquine is known to increase the excretion of uroporphyrins in the urine, we present a patient with a good response to low dose hydroxychloroquine.

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Cell-free autologous conditioned serum (ACS) improves skin elasticity: Results of an investigator initiated clinical trial OrthoSkin I

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Background: The therapeutic safety and efficacy of ACS has been shown in controlled clinical trials for knee osteoarthritis and lumbar radiculitis. ACS's efficacy has been ascribed to high levels of cytokines and growth factors (eg, TGF-β, IGF1), achieved through incubation, which distinguishes ACS from PRP. Increase of collagen type I has been shown histologically in animals treated with ACS. Skin damage due to extrinsic factors increases oxidative stress, which leads to reduction of TGF-β levels and collagen synthesis in skin. Substitution with autologous TGF-β in ACS may increase skin elasticity and have rejuvenative effects.

Methods: 21 female patients (age 35-55) with reduced facial skin elasticity have been treated via microneedling with cell-free ACS in three sessions at day 0, week (W) 2 and W4 (by inefficacy additionally at W12) in an open, monocentric, prospective 24W clinical trial approved by the ethical committee. The primary endpoint was skin firmness and elasticity measured by Cutometer after 12W and 24W. Validated questionnaires and Corneometer were secondary parameters, all measurements were performed under standardized conditions. Safety has been evaluated.

Results: All cutometer parameters for elasticity (R0-R9) improved statistical significantly by W12 and W24. Improvement in R0 (passive behavior of the skin to force) was from 0.34 to 0.24 and in R5 (net elasticity) from 0.36 to 0.45 (29% respectively 25%, both $P < .01$). The results in GAIS questionnaire correlated with quantitative skin measurements (improvement of 59.5% in investigator's and 57.1% in patient's estimation at W12). The skin hydration increased from 42.61 to 54.70 within 12W (28.3%, $P < .01$). No adverse events were observed.

Conclusions: This study for the first time reveals a significant improvement of cutaneous elasticity and skin firmness thus leading to a visual and measurable gain of rejuvenation. ACS has the potential to be the first cell-free biological skin enhancer from patient's own blood. Further clinical investigations are ongoing for better understanding of the underlying mechanisms.

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Case report: Squamous cell carcinomas arising in acral with chronic arsinicism and psoriasis successfully treated with insulin, zinc hyaluronate after surgically excision

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Squamous cell carcinomas (SCC) is rarely associated with psoriasis and chronic arsinicism. Healing of this SCC after excision is very difficult because of performing "skin graft" on the hyperkeratosis lesion caused by chronic arsinicism, especially at the distant acral sites. Treated with insulin, zinc hyaluronate after surgically excision is very successful. We report a case of a 46 -year-old male with a 17-year history of poorly controlled psoriasis and 4 years of traditional medicine taken presented with two indolent ulcerative lesions at widely distant acral sites of the right ventral hand and the right calcaneal sole. The lesions were histologically confirmed as squamous cell carcinomas and surgically excised. The other lesion sites were found and excised. After excision, the wounds didn't heal especially at the right hand and right calcaneal sole. Examination showed hyperpigmentation and hyperkeratosis of the left palmar skin, a rain-drop pattern of hypopigmentation in both axillae. Each of these features could have been consistent with underlying psoriatic skin disease. Arsenic was present in the hair sample at a concentration of 2.15 mg/kg. However, after excision, the wounds didn't heal especially at the right hand and right calcaneal sole. These are also the special sites that are difficult to perform "skin graft." Then, we treated the patient with topical insulin and zinc hyaluronate at these areas twice a day. After 8 weeks, the lesions were completely healed. Thus, this new treatment showed the effectiveness on the healing process, especially for severe lesion cases.

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Central facial inflammation: A manifestation of nickel allergy

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Nickel is the most common allergen in patch test clinics worldwide. Oral ingestion of nickel-containing foods is known to cause manifestations of systemic contact dermatitis in sensitive individuals including eczematous dermatitis, pruritus ani and labial lichen planus. We present a case of systemic contact dermatitis due to dietary nickel presenting as an inflammatory process mimicking rosacea. A 59-year-old white female presented with a 4-month history of a tender, erythematous, papular eruption on her forehead, chin and bilateral cheeks. She was previously diagnosed with rosacea, but failed to respond to standard treatment including doxycycline, azelaic acid and tretinoin. She noted no specific exacerbating or alleviating factors, but noted that the eruption looked worse at the end of the day despite avoiding common rosacea triggers including UV, alcohol and spicy foods. Her past medical history was remarkable only for earlobe dermatitis as a teen when she started wearing jewelry. The patient was given a diagnosis of severe inflammatory rosacea and treated with dexamethasone and cephalexin. The eruption initially cleared, then recurred. Again, it responded to dexamethasone and cephalexin. Since she improved with steroid, she underwent patch testing which revealed a non-vesicular reaction to balsam of Peru, nickel sulfate, bronopol and thimerosal. Patient denied using products with these preservatives or fragrances, and had not been vaccinated within the previous 5 years. Of note, the patient reports that one month prior to onset she began a strict vegetarian diet, which is known to be high in nickel. In retrospect, she recalled exacerbation by ingestion of herbal teas. Patient began a nickel-low diet by avoiding whole wheat, beans, cocoa, nuts, canned foods and shellfish. She also began daily vitamin C, which enhances iron absorption, and increased her intake of iron, a competitive inhibitor of nickel absorption. With this diet, her facial eruption improved dramatically within 5 weeks and completely resolved in 5 months. A challenge with her previous diet reproduced the facial inflammatory dermatosis. Given her improvement with the diet, the patient likely had a systemic contact dermatitis mimicking inflammatory rosacea due to dietary nickel. This case serves as an example to consider nickel allergy in patients who fail therapy for other dermatologic conditions.

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