Positive HLA Helps Shed Light on Actinic Prurigo

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PORTLAND, ORE. — Actinic prurigo, a disorder once considered a variant of polymorphous light eruption in Native Americans, may be neither a form of PMLE nor a condition limited to Native Americans, Dr. Lisa H. Williams said at the annual meeting of the Pacific Northwest Dermatological Society.

"Classically described in Native Americans, actinic prurigo is now increasingly recognized in [white] populations," said Dr. Williams, a dermatology resident at the University of Washington in Seattle.

Actinic prurigo falls within the category of idiopathic photosensitive dermatoses, but it has several distinguishing characteristics. Its onset is usually within the first decade of life and it may improve by puberty. In addition, it can be tricky to trace outbreaks to sunlight, because lesions are persistent and may occur on sun-exposed or non–sun-exposed skin at any time of the year.

Dr. Williams described two patients, the first, a 6-year-old white boy with a 2-year history of excoriated papules that appeared year-round in contrast to the seasonal appearance of most PMLE eruptions. The lesions appeared on his forehead, cheeks, ears, lower back, calves, and shins. He was otherwise a healthy child with no personal or family history of atopic dermatitis. Phototesting yielded normal findings.

Pivotal in the diagnosis of the boy was a positive association with an HLA DR4 test subtype: DRB1*0407. The test is positive in 60%-70% of white patients with actinic prurigo and in 4%-8% of controls. The less specific HLA type DR4 is seen in 82%-96% of patients with actinic prurigo and in 30%-40% of people who do not have the disorder.

Similarly, Mexican mestizos with actinic prurigo are likely to test positive to HLA A29-B39, and Canadian Cree First Nations people tend to test positive to the HLA A24 subtype.

"There are different HLA types for each different ethnic group that can have the disorder. [It] can really help you when you're trying to diagnose it," Dr. Williams said.

Native Americans are, in fact, more likely than are other patients with actinic prurigo to experience conjunctivis and/or cheilitis, which can be excellent diagnostic clues. Conjunctivitis occurs in about 10%-30% of all patients.

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A second case featured by Dr. Williams was a 7-year-old Mexican American boy with both conjunctivitis and cheilitis. These were key to the diagnosis of actinic prurigo, along with the fact that his photosensitive rash consisted of itchy papules that developed 30 minutes after sun exposure.

The lack of vesicles, burning pain, crusting, scarring, or an elevated red blood cell protoporphyrin test ruled out other differential diagnoses, including hydroa vacciniforme and erythropoietic protoporphyria.

Dr. Williams said recent publications call into question the traditional notion that actinic prurigo is a form of PMLE. Clinically, there are sharp differences, including the fact that PMLE is noteworthy for "hardening," a sparing of the face and the dorsal hands after repeated exposure to the sun. Clothing-covered areas are rarely involved in PMLE, in contrast to actinic prurigo.

Phototesting is more sensitive in actinic prurigo, producing positive results in about 60% of patients, compared with only about 20% of patients with PMLE.

In the case of the 6-year-old, the positive HLA subtype pointed solidly to actinic prurigo. His parents kept him inside during a 2-week winter school vacation and he cleared.

Both children are being treated with antimalarial drugs.

Other treatment options include psoralen ultraviolet light treatment, corticosteroids, antihistamines, vitamin E, pentoxifylline, β-carotene, and especially thalidomide. The latter is so effective for the treatment of actinic prurigo that some investigators believe that a trial of the drug, if successful, could nail down the diagnosis. ■