# Vitiligo May Be First Sign of Localized Scleroderma

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CHICAGO — When facing a child with localized scleroderma, be wary if the scleroderma is linear, especially if it is on the face or on a limb where it crosses a joint, Dr. Amy Gilliam said at the annual meeting of the Society for Pediatric Dermatology.

However, not all young people initially present with the skin hardening and atrophy that characterizes scleroderma, explained Dr. Gilliam, a dermatologist at the University of California, San Francisco. Instead, these patients are often misdiagnosed with vitiligo for months or years before the correct diagnosis of juvenile localized scleroderma is made.

To better characterize localized scleroderma in children, Dr. Gilliam and her colleagues reviewed data from 127 patients younger than 21 years who were evaluated at UCSF. Dr. Gilliam's research was supported in part by a grant from the Society for Pediatric Dermatology and the Dermatology Foundation, but she had no other financial disclosures.

"Something unique in our study is that we collected information on body surface area of involvement," Dr. Gilliam said. "And we had a dermatology perspective rather than a rheumatology perspective.

"One of the interesting things that came out of our data was that the presenting sign in about 50% of the patients was some type of dyspigmentation, either hyper-, hypo- or depigmentation," she said. Add the 19 patients who had what they called a "bruise," and dyspigmentation was a presenting symptom in nearly two-thirds of the cases.

Another key finding was that patients whose scleroderma involved 5% or more of their total body surface area were significantly more likely to have extracutaneous symptoms—including arthralgias and orthopedic, pulmonary, and gastrointestinal problems—than were patients whose scleroderma involved less than 5% of their total body surface area. The significance was true in separate analyses of the 89 patients whose charts were reviewed retrospectively and the 38 patients who were studied prospectively and followed.

But neurologic problems were the notable exception in the patient population. "That sticks out like a sore thumb," said Dr. Gilliam. Localized scleroderma on less than 5% of the body surface area was significantly associated with neurologic problems, and neurologic problems were significantly more common in patients with facial linear scleroderma.

This finding "makes complete sense to me, because when we are talking about neurologic problems in the setting of localized scleroderma, we are usually talking about the face, which has at most 6% of the surface area, so these patients with neurologic problems are likely to have lower total body surface area involvement," she said.

Apart from the relationship with body surface area, Dr. Gilliam was able to prove that neurologic problems were more common in patients with facial linear scleroderma compared with those who had other forms of localized scleroderma (33% vs. 8%). Her data also showed that orthopedic problems were significantly more common in patients with nonfacial linear scleroderma, compared with those who had other forms of localized scleroderma (22% vs. 2%).

But body surface area alone is not enough to assess localized scleroderma, Dr. Gilliam emphasized. The patients to worry about are those with segmental or linear presentations and those with the characteristic pinkish-purple macules that indicate generalized morphea.

It's important to think about location in cases of localized scleroderma, Dr. Gilliam added. In her study, gastrointestinal problems were significantly more common in patients with generalized morphea and in patients who had scleroderma on the trunk, compared with those who had scleroderma in other locations (21% vs. 5%). But location isn't everything: Pulmonary problems were significantly more common among patients with generalized morphea, but the presence or absence of localized scleroderma on the trunk was not significant.

Lastly, Dr. Gilliam did not find a significant association between positive levels of antinuclear antibodies and extracutaneous conditions, although she cited a separate study of 750 patients that did show a significant association (Arthritis Rheum. 2005;52:2873-81). ■