Dyspigmentation May Mean Localized Scleroderma

BY HEIDI SPLETE
Senior Writer

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

Not all young people initially present with the skin hardening and atrophy that characterizes scleroderma, explained Dr. Gilliam, of the University of California, San Francisco. 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 under 21 years 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. "We collected information on body surface area of involvement. And we had a dermatology perspective rather than a rheumatology perspective," Dr. Gilliam said.

"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 finding was that patients whose scleroderma involved 5% or more of total body surface area were significantly more likely to have extracutaneous symptoms—arthralgias and orthopedic, pulmonary, and gastrointestinal problems—than were patients whose scleroderma involved less than 5% of total body surface. This held in separate analyses of 89 patients whose charts were reviewed retrospectively and 38 patients 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.

"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 showed neurologic problems were more common in patients with facial linear scleroderma versus other forms of localized scleroderma (33% vs. 8%). Her data 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 isn't enough to assess localized scleroderma, Dr. Gilliam said. 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.

Dr. Gilliam did not find a significant link 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). She found positive antinuclear antibody levels were slightly, but not significantly, more prevalent in patients with linear scleroderma and with generalized morphea. ■

Long-Term Etanercept Use in Children With RA Appears Safe

BY JEFF EVANS
Senior Writer

Etanercept appears safe through 8 years of continuous use in children with polyarticular-course juvenile rheumatoid arthritis without any incidence of cancers or serious opportunistic infections.

That is the conclusion of a multicenter, randomized, controlled trial, later extended into an open-label study, looking at the long-term safety of the anti–tumor necrosis factor–α drug. "Etanercept [Enbrel] was the first of the anti-TNF agents tested in children with JRA, and thus we have the longest and most clinical experience with it," said Dr. Daniel Lovell of the Cincinnati Children's Hospital Medical Center, in an interview.

Nonimmune children on etanercept should not be exposed to varicella because of increased susceptibility.

DR. LOVELL

Of 69 patients originally randomized into the trial, Dr. Lovell and Dr. Andreas Reiff and colleagues followed 58 into the open-label extension; all had taken at least one etanercept dose.

In the original trial, all patients initially received etanercept for 3 months. Those who responded were randomized to etanercept or placebo. Corticosteroids and anti-inflammatory drugs were allowed in both the randomized trial and the extension, but few patients were allowed to take methotrexate again, said Dr. Reiff, head of the division of rheumatology and rehabilitation at Children's Hospital Los Angeles. In the ongoing safety study, 42 of the original patients (61%) received at least 4 years of etanercept, and 16 (23%) received at least 8 continuous years of the drug. All used the recommended dosage for the subcutaneous injection formulation (0.4 mg/kg twice per week). Overall, 16 patients (23%) reported adverse events. Long-term use did not significantly increase the rate of adverse events. Between years 4 and 8 of follow-up, only one severe adverse event (pyelonephritis) occurred. No lupus, demyelinating disorders, tuberculosis, opportunistic infections, malignancy, or deaths were reported.

Three cases of varicella infection occurred in follow-up, according to Dr. Reiff. Although Dr. Lovell said etanercept had a good safety profile, he expressed caution about an increased risk of varicella in children on etanercept who lack protective antibodies to this virus.

Dr. Lovell recommended all children be tested for varicella antibodies (IgG) before etanercept treatment. Precautions should be taken to avoid exposing nonimmune children on etanercept to active varicella.

Even patients on immunosuppressive drugs who were vaccinated may not have a protective antibody level, said Dr. Reiff, who noted he serves as an adviser and speaker for Amgen Inc. and Wyeth, which comarket etanercept.

"There is definitely a possibility that varicella can be more severe on these drugs," Dr. Reiff said in an interview. But since etanercept has a half-life of nearly 5 days, it takes 3-4 weeks before it is cleared from the body after stopping. "We watch these patients very carefully. We treat them for their chicken pox, and if we see that their course is aggressive or prolonged, we admit them for IV treatment."

Dr. Lovell noted when parents vaccinate or revaccinate nonimmune children against varicella, initiation of etanercept is delayed by 3 months. Prophylactic acyclovir is needed in children on etanercept with known exposure to varicella. ■