Pediatric Facial Eruptions Flag Inadequate Nutrition

The typical rash associated with poor diet doesn't itch and may not respond to topical steroids.

BY JANE SALODOF MACNEIL
Southwest Bureau

PARK CITY, UTAH — Physicians should consider nutritional deficiencies when diagnosing facial eruptions in infants and children, according to Dr. Beth Drolet.

One of the more perplexing cases she described at a clinical dermatology seminar sponsored by Medicis was a 2½-year-old child referred by emergency physicians for suspected Stevens-Johnson syndrome.

Dr. Drolet, medical director for dermatology at Children's Hospital of Wisconsin, Milwaukee, said the child had no history of health care or immunizations. He was brought to the hospital because of a severe rash and was found to have multiple deficiencies, including severe sensory polyneuropathy, photophobia, muscular atrophy, osteopenia, and speech and language defects.

"The child was only eating large french fries from McDonald's. He was getting enough calories, but not enough vitamins," she said, reporting the parents said that was all he would eat. Although the boy's diet has been corrected, she reported he still has severe neuropathy and mental delay.

In another pediatric case, Dr. Drolet said that toxic epidermal necrolysis was suspected in a child with an "extremely smelly, flaky eruption." It turned out the child had been diagnosed with a milk allergy and his diet was almost entirely Rice Dream, a nondairy beverage touted as a substitute for dairy milk.

Although Rice Dream is enriched with vitamins A, D, and B_{12} and has comparable calcium to dairy milk, it provides little protein. Indeed, its label warns that it should not be used for infant formula or in children under 5 years of age without consulting a physician.

"This [Rice Dream] is not a bad thing if the child is getting other nutrition," said Dr. Drolet, also of the Medical College of Wisconsin, Milwaukee.

Dr. Mark Davis of the Mayo Clinic in Rochester, Minn., described a similar case in a separate presentation on hospital dermatology at the meeting. In that case, a 2-year-old boy with rash and hair loss was diagnosed with kwashiorkor. This child also was on a Rice Dream diet, according to Dr. Davis, who emphasized the importance of history in making a diagnosis.

If a child has a nutritional deficiency, Dr. Drolet said, the facial eruption could be described by the following morphology: large plaques; large, thin scales; sharply demarcated, irregular, driplike borders; and superficial erosions.

The eruption is not itchy, she said, and it does not respond to topical steroids. The distribution is periorificial and widespread.

In addition, the child may have lackluster, hypopigmented hair with the "flag sign"—a band of lighter hair associated with nutritional deficiency.

"How does this happen in developed countries in 2006?" Dr. Drolet asked rhetorically, answering her own question with "real/perceived milk allergy, fad diets, behavioral problems, food preferences, [and] nutritional ignorance."

Among the other nondermatologic causes of facial eruptions that could be considered when making a diagnosis, Dr. Drolet cited herpes, pediatric Horner syndrome, staphylococcus infection, juvenile idiopathic arthritis, neonatal lupus, irritable bowel disease, Crohn's disease, Henoch-Schönlein purpura, drug-induced hypersensitivity syndrome, dermoid cyst, and PHACE (posterior fossa brain malformations, hemangiomas of the face, arterial anomalies, cardiac anomalies, and eye abnormalities) syndrome. ■

Recognize, Aggressively Treat Cutaneous Evidence of Lupus

BY DIANA MAHONEY
New England Bureau

STOWE, VT. — Skin involvement is one of the most frequent manifestations of lupus erythematosus, yet the cutaneous signs of the disease are not always recognized, Dr. Victoria P. Werth said at a dermatology conference sponsored by the University of Vermont.

The diagnostic challenge is recognizing these cutaneous conditions, particularly because they encompass as many as 15 different clinical presentations, said Dr. Werth of the University of Pennsylvania, Philadelphia.

It's important to know what to look for, "not only so we can treat the skin problems, but also so we can identify signs of systemic disease when they occur," she said.

Although systemic lupus erythematosus (SLE) is estimated to occur in 17-48 per 100,000 individuals, the cutaneous variants are thought to be two to three times more prevalent.

Skin findings in cutaneous lupus are generally categorized into lupus-specific and lupus-nonspecific diseases, based on biopsy findings.

"Lupus-specific lesions show histology that is specific to lupus erythematosus, while nonspecific lesions are not histopathologically distinct for the disease and may be seen as a feature of another disease process," Dr. Werth explained. Some of the more common nonspecific skin findings include alopecia, vasculitis, and Raynaud's phenomenon.

Although skin disease is the second-most frequent clinical manifestation of SLE, cutaneous lupus does not always meet all the American College of Rheumatology's diagnostic criteria for SLE.

Lupus-specific cutaneous lesions are further subdivided into three categories: acute, subacute, and chronic. "The most recognizable acute presentation is the butterfly rash, which comes on abruptly and heals within hours or days, usually without scarring," Dr. Werth said. Some variations of this rash include bullous formations or blisters.

Subacute manifestations can include annular and/or psoriasiform rashes that are usually highly photosensitive. Chronic cutaneous lupus is "the wastebasket category" for many of the other lupus-specific skin presentations, she said.

The most common chronic cutaneous form is discoid lupus erythematosus, which begins with well-defined scaly lesions that evolve into scarring plaques and often includes follicular involvement that can lead to hair loss.

"Early, aggressive treatment for these patients is important in order to prevent permanent, disfiguring scarring and permanent hair loss," said Dr. Werth.

She also discussed several of the following less prevalent manifestations of chronic cutaneous lupus:

▶ **Chilblain lupus.** Associated with itching, cold, and painful swelling of the extremities and toes.

▶ **Hypertrophic lupus.** Characterized by wartlike bumps.

▶ **Lupus profundus.** Causes deep dermal nodules on the upper arms and sometimes on the head, chest, or legs.

▶ **Lupus tumidus.** Presents as broad, indurated plaques.

Because some of these conditions can mimic other skin conditions, "serologic and histologic examinations are critical for confirming a diagnosis," Dr. Werth said.

Although management for the various lupus subsets does not differ substantially, the clinical distinctions are important because they relate to the likelihood that an individual patient will develop systemic disease, Dr. Werth pointed out.

There's little in the way of literature. Often, the presumed efficacy of the available agents has been extrapolated from their use in systemic lupus.

Nearly all patients with acute cutaneous lupus, and half of those with subacute disease, will meet American College of Rheumatology criteria for SLE. With respect to chronic cutaneous lupus, those with generalized discoid lesions have a 20% chance of developing SLE, those with lupus profundus are estimated to have a 10% chance, those with localized discoid lesions have a 5% chance, and those with lupus tumidus have virtually no chance.

"Patients whose skin conditions put them at higher risk should be followed more closely for evidence of systemic disease," she said.

Once a diagnosis of cutaneous lupus has been made, the next step is to evaluate the patient for signs of systemic disease. The initial evaluation should include history, physical examination, CBC, sedimentation rate, antinuclear antibody (ANA) testing, and urinalysis. For ANA-positive patients or ANA-negative patients in whom there is suspicion of SLE, Dr. Werth suggests a panel that includes anti-Sjögren's syndrome A, anti-Sjögren's syndrome B, anti-ribonucleoprotein, anti-double-stranded DNA, and anti-Smith antibodies, as well as complement studies.

Patients with positive findings warrant closer monitoring, "especially those with high titer, anti-double-stranded DNA antibody because of an increased risk for renal problems," she said.

In terms of management, "the first order of business is advising patients to avoid precipitating factors such as heat, certain medications, and sunlight," Dr. Werth said. "I tell my patients they should not go outside without a sunscreen that has a UVB of 30 or more, as well as a UVA blocker, like Parsol 1789."

Patients also should avoid drugs that cause subacute lupus-like eruptions, such as thiazides, griseofulvin, terbinafine, calcium channel blockers, and ACE inhibitors, Dr. Werth noted.

In addition to preventive efforts, standard therapies include topical and sometimes intralesional corticosteroids and antimalarial drugs.

"Most patients will respond well to these conservative therapies, but there are 10%-20% of patients with refractory disease who don't respond," she said. For such patients, there are a range of other treatment options, including thalidomide, dapsone, retinoids, biologics, and immunosuppressants.

"Some of these more aggressive therapies are associated with significant side effects, and should be used sparingly and only when warranted by disease severity," Dr. Werth said.

"Unfortunately, there is a paucity of data in the literature to aid the clinician in terms of which therapies to employ. Often, the presumed efficacy of these drugs has often been extrapolated from their use in treating systemic lupus."

It is hoped that new insights into the genetic pathways associated with cutaneous lupus, as well as continued advances in targeted biologic therapies, will lead to the development of more selective, less toxic therapeutic agents.

Among the drugs currently in development, Dr. Werth said, "are a derivative of thalidomide and a new B-cell-specific monoclonal antibody therapy." ■