Pediatric Pearls—From Langerhans to Kawasaki

Consider Langerhans cell histiocytosis in children with refractory atopic or seborrheic dermatitis.

BY BRUCE JANCIN
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WAIKOLOA, HAWAII — Consider Langerhans cell histiocytosis in the child with crusted papules on the palms and soles who has been unsuccessfully treated for scabies or who is mite negative under the microscope, a pediatric dermatology expert suggested.

The diagnosis of Langerhans cell histiocytosis is often delayed for months, sometimes even more than a year, with the lengthiest delays occurring in infants and toddlers. A common reason for the delay is a misdiagnosis of scabies, Dr. Anthony J. Mancini said at the annual Hawaii dermatology seminar sponsored by Skin Disease Education Foundation.

Another reason for delayed diagnosis is that the clinical presentation of Langerhans cell histiocytosis in neonates can differ from that in older children, which is a problem because the disease is more common among neonates than are varicella and neonatal herpes.

Neonates with Langerhans cell histiocytosis can present with hemorrhagic vesiculopustular lesions rather than with the classic scaly, erythematous lesions that occur in older children, according to Dr. Mancini, head of pediatric dermatology at Children's Memorial Hospital, Chicago.

He offered up the following selection of pediatric dermatology clinical pearls, from Langerhans cell histiocytosis to Kawasaki disease:

► **Langerhans cell histiocytosis.** Formerly known as histiocytosis X, this disorder ranges in severity from a benign and self-limited condition to a disseminated disease with severe morbidity and even mortality. The skin and bones are the most commonly involved sites. Other sites include the GI tract, lungs, spleen, CNS, liver, and endocrine glands.

A common theme of Langerhans cell histiocytosis in the skin is discrete superficial papular or nodular erosions in flexural folds. The papules are brown to red in color and may be flat topped, especially in infants. Consider the possibility of Langerhans cell histiocytosis in children with refractory atopic or seborrheic dermatitis.

"Don't hesitate to get a biopsy," Dr. Mancini urged.

► **Streptococcal intertrigo.** Think of group A β-hemolytic streptococci in children with fiery-red intertrigo with a foul smell, erosive lesions, and an absence of the satellite lesions typical of *Candida albicans* intertrigo.

Streptococcal intertrigo responds quickly to treatment with a low-potency, topical corticosteroid for the inflammation, moist compresses two or three times per day, perhaps a topical antifungal agent to address the *Candida*, and, most importantly, a systemic antibiotic that covers both group A β-hemolytic streptococci and possible coinfection with *Staphylococcus aureus*.

"Your first choice here should be something simple, like cephalexin given t.i.d. for 10 days," said Dr. Mancini.

► **Serum sickness–like reactions.** These drug reactions can occur 1-3 weeks after starting any of a number of antibiotics or griseofulvin, and are not true serum sickness because there are no circulating immune complexes. Fever and swollen lymph nodes occur less often than in true serum sickness, and proteinuria is rare.

The cutaneous hallmark of serum sickness–like reactions is erythema progressing to purple urticaria with dusky centers. The lesions tend to be transient and migratory. The child will have periarticular swelling, arthralgia, and a reduced range of motion. Refusal to ambulate is a key feature.

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DR. MANCINI

Treatment consists of the withdrawal of the offending drug and a prescription for a nonsteroidal anti-inflammatory agent and for corticosteroids, Dr. Mancini said at the meeting.

► **Herpes-associated erythema multiforme.** Don't hesitate to suppress this disease, both to prevent the painful and unsightly lesions of erythema multiforme and to head off mucous membrane involvement, and because treatment may prevent Stevens-Johnson syndrome.

Acyclovir gets the nod here because it is approved for use in children; famciclovir and valacyclovir are not. "I like the acyclovir oral suspension; it's really well tolerated in kids," Dr. Mancini said. He prescribes ½ teaspoon (100 mg) twice daily in children younger than 2 years old, 150 mg twice daily in children aged 2-6 years, and a full teaspoon twice daily in those older than 6 years.

The appropriate duration of therapy to suppress herpes-associated erythema multiforme is unclear. He recommends a minimum of 6 months of daily acyclovir, then a taper to a trial off therapy. In some cases, treatment needs to continue for years. "This is an art, not a science," the dermatologist stressed.

► **Kawasaki disease.** This disease is the leading cause of acquired heart disease in patients under 5 years of age. Formal diagnosis requires the presence of any five of these six symptoms: unexplained high fever lasting more than 5 days; cervical adenopathy in excess of 1.5 cm; nonpurulent conjunctival infection; swelling of the extremities; oropharyngeal changes; and a polymorphous skin eruption.

Coronary artery aneurysms also can occur with incomplete Kawasaki disease, marked by high fever plus as few as one or two of the other criteria. This occurs more commonly in infants.

The cutaneous finding of perineal accentuation or desquamation should increase concerns about the possibility of Kawasaki disease. In contrast, there is much less worry about Kawasaki disease in a child with blisters or purpura.

Dr. Mancini had no conflicts of interest to disclose.

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Nickel Adding to Contact Dermatitis Diagnosis Conundrum

BY CAROLINE HELWICK
Contributing Writer

NEW ORLEANS — Increases in nickel allergy have done nothing to make the diagnosis of contact dermatitis a more exact science, according to a pediatric dermatologist.

In her update on contact dermatitis, Dr. Lisa Garner, a private practitioner from Garland, Tex., also discussed the diagnosis of fragrance, local anesthetic, and plant allergies.

Nickel recently received special recognition from the American Contact Dermatitis Society ("Nickel Named Contact Allergen of the Year," SKIN & ALLERGY NEWS, February 2008, p. 1).

It is the most frequent positive patch test allergen worldwide, and in the United States only poison ivy / oak is responsible for more allergic contact dermatitis (ACD). Nickel allergy now stands at almost 19% of all persons who are patch tested in referral centers, though the definite relevance is far less, Dr. Garner said at a dermatology update sponsored by Tulane University.

Body piercing appears to be the primary risk factor for ACD today, while nickel in earrings, snaps, and belt buckles remains problematic. In one study, investigators found that 16% of blue jeans tested positive for nickel (Dermatitis 2007;18:208-11). Coating these products with a nail hardener or a nickel guard can prevent the release of nickel.

There are now reports of reactions to orthopedic devices, orthodontic appliances, and, more recently, stents, with anecdotal reports of restenosis occurring more frequently in nickel-allergic patients. "But if you see dermatitis over a knee implant, this is more likely to be a reaction to an analgesic rub, tea tree oil, or other substance. First look for these as a cause of the local reaction," she advised.

Nickel-allergic patients can assess the nickel content of household and personal objects using the Delasco or Allertest Ni (Allerderm) kits.

Fragrance was the American Contact Dermatitis Society's allergen of the year in 2007 (Dermatitis 2007;18:3-7) and is the fourth most common patch test, though definite relevance is hard to document for an individual patient, Dr. Garner said.

Fragrances are practically ubiquitous in products, and individual compounds are not listed on product labels. "Having an allergic reaction to one fragrance does not mean a patient will develop ACD to all fragrances, and there is no way to find out which fragrance a patient is actually allergic to," she said. "Patients with a strong reaction to patch testing will have to be cautious with the introduction of any new product. The stronger their reaction, the more likely they are to be strongly allergic to another fragrance."

To improve the ability to detect fragrance allergy, six European centers developed the fragrance mix II patch test in 2005. This test contains citronellol, lyral, hexyl cinnamal, citral, coumarin, and farnesol.

Still rare, though becoming more common, is ACD to local anesthetics, since these compounds are being increasingly included in prescription and over-the-counter products. Allergic reactions include localized or generalized eruptions as well as delayed-type hypersensitivity reactions, said Dr. Garner, who had no conflicts to disclose.

In 2007, investigators reported 16 cases of positive patch test reactions to lidocaine out of 1,143 patients tested over 5 years (Dermatitis 2007;18:215-20). Two had lidocaine as their only positive test, and three of eight who underwent intradermal testing had positive reactions.

Certain natural compounds are responsible for an increasing number of positive patch tests. Toxicodendron, Compositae, and tea tree oil are the most common botanic allergens; they contain sesquiterpene lactones, over 100 of which have been identified, she said.

Patients can be tested using the sesquiterpene lactone mix (alantolactone, dehydrocostus lactone, and costunolide), which is specific but lacks sensitivity, and the compositae mix (short ether extracts of arnica, Germany chamomile, yarrow, tansy, and feverfew flower extra), which is more sensitive. Tea tree oil elicits positive patch testing in 0.5% of individuals, so it has been added to the North American Contact Dermatitis Group standard tray.

"Be sure to patch test the patient to their own products," Dr. Garner added.

Dr. Garner also noted that new thin-layer rapid-use epicutaneous test allergens were released this year, including imidazolidinyl urea, diazolidinyl urea, tixocortol-21-pivalate, and budesonide. Patch testing to tixocortol and budesonide identifies 91% of steroid-allergic patients (including D2 steroid patients) but does not identify patients allergic to betamethasone valerate or clobetasol propionate (D1 steroids). Quinolone mix has also been added, which should identify allergen sources in paste bandages, topical antibiotics and antifungal creams, lotions and ointments, and animal food. ■