Puzzling Out Difficult Pediatric Skin Diagnoses

BY HEIDI SPLETE
Senior Writer

ORLANDO — Pediatric skin conditions often pose diagnostic challenges because many cutaneous disorders have similar clinical features.

Annular lesions of granuloma annulare may be mistaken for tinea corporis; follicular papules of keratosis pilaris may be confused with follicular eczema; and nail psoriasis may be misdiagnosed as onychomycosis.

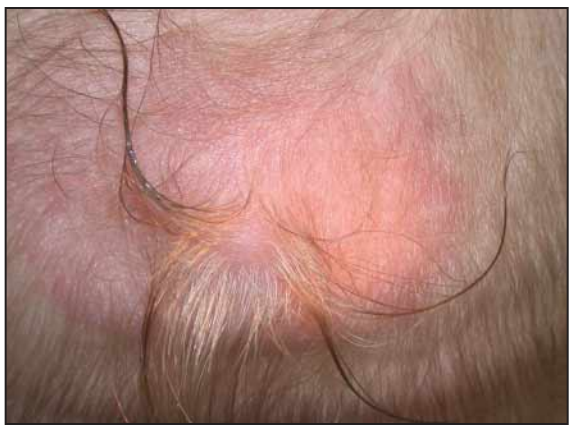
At a meeting sponsored by the American Academy of Pediatrics, Albert C. Yan, M.D., director of pediatric dermatology at the Children's Hospital of Philadelphia, provided some helpful diagnostic tips for distinguishing some of these potentially puzzling skin problems.

The Hair Collar Sign

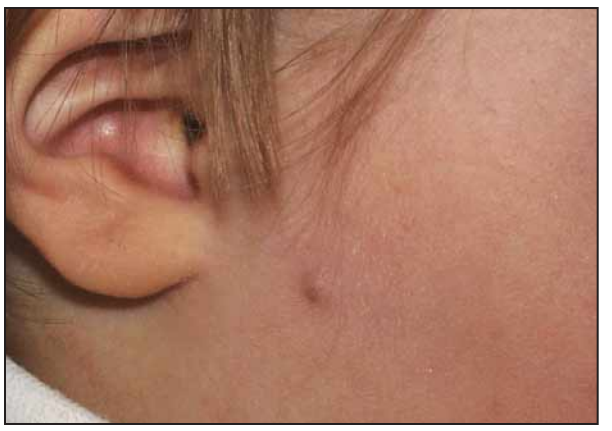
A boy is born with an area of localized, circular alopecia covered by a glossy membrane. The area is surrounded by a collection of dark, terminal hairs. Palpation reveals that a lump is present.

Occasionally mistaken for fetal scalp monitor trauma, neonatal herpes simplex infection, or a nevus sebaceus of Jadassohn, this characteristic pattern—a collar of coarse hair surrounding an area of membranous aplasia cutis congenita—can be a marker for cranial dysraphism, a developmental defect of the skull potentially associated with structural neurologic defects. The scalp defect may represent only the tip of the iceberg, Dr. Yan noted, since underlying bony defects or ectopic brain tissue may be present.

In such cases, magnetic resonance imaging is essential to rule out underlying abnormalities, including atretic encephaloceles or heterotopic brain tissue.



A collar of hair around membranous aplasia cutis congenita can indicate cranial dysraphism.



Pilomatricomas may be associated with systematic disorders such as Gardner's syndrome or sarcoidosis.

Pilomatricoma

A 15-month-old girl presented with a bump on her cheek—a firm, bluish, cystic papule that moved back and forth under pressure. When one end of the lesion was palpated, the other end would pop up, a phenomenon also known as a "teeter-totter sign."

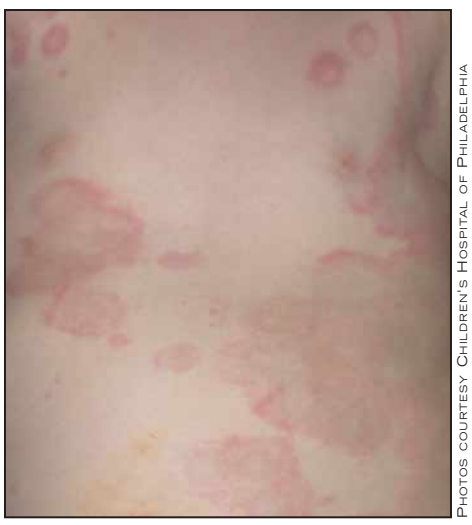
Although these lesions may resemble dermoid cysts or epidermal inclusion cysts, the diagnosis in this case was pilomatricoma, distinguished by its bluish color and the presence of the teeter-totter sign. The lesions most often occur on the head or neck, although other areas occasionally are affected. Pilomatricomas generally are solitary, benign, frequently calcified, and arise from hair follicles. In some cases, the lesions resolve spontaneously, but more often, they persist and grow, and surgical intervention is recommended. Pilomatricomas may rupture, which can cause inflammation and scarring. Although pilomatricomas generally are isolated findings, they may be associated with systemic disorders such as Gardner's syndrome, myotonic dystrophy, and sarcoidosis.

Annular Urticaria/Urticaria Multiforme

A 3-year-old girl presented with red, swollen, annular plaques on her skin, and had swollen hands and feet. She had been otherwise healthy and was taking no medications. The condition arose suddenly; the parents noticed the rings and swelling one morning when picking up the child from her bed. On closer inspection, the rings were red and blanchable, with clear white centers. Some were imperfect circles.

The diagnosis is annular urticaria. "These types of cases are frequently referred for suspected erythema multiforme," Dr. Yan noted. "Lesions of annular urticaria are evanescent; the lesions fade and move, and the lesions can form imperfect circles with clear centers. The lesions may disappear within 24 hours, only to show up elsewhere," he said. By contrast, erythema multiforme appears as fixed target, or "bull's-eye," lesions with dusky centers and is associated with mucous membrane ulcers.

The two conditions are treated quite differently, Dr. Yan emphasized. Annular urticaria responds to combinations of antihistamines or occasionally steroids; erythema multiforme requires a detailed history to determine underlying causes, removal or treatment of those causes, and consideration of steroid therapy if indicated. Dr. Yan often refers to annular urticaria as "urticaria multiforme" because these cases are so regularly mistaken. ■



PHOTOS COURTESY CHILDREN'S HOSPITAL OF PHILADELPHIA

Urticaria multiforme, often mistaken for annular urticaria, is linked to allergy.

Early, Aggressive Surgery Is Best Course for Neurofibroma in Children

BY PATRICE WENDLING
Chicago Bureau

PARIS — Resection can't be too early or too aggressive when treating neurofibroma in children, reported McKay McKinnon, M.D., at the Fourth International Academy of Cosmetic Dermatology World Congress.

Neurofibroma is a usually benign tumor of the peripheral nerves caused by abnormal proliferation of Schwann cells.

The development of neurofibromas, multiple café au lait spots, and freckling in the armpits or groin area are common symptoms of neurofibromatosis 1 (NF1), which occurs in 1 in 4,000 births.

There is no way to tell which patients will develop a mild case and which patients will have serious complications such as painful or disfiguring tumors.

Tumors may grow back and in greater numbers, and they could be fibrosarcoma, said Dr. McKinnon, a plastic and reconstructive surgeon with the University of Chicago. Early surgery, even in infancy, may be indicated.

"In my experience, a very aggressive surgery can often produce nonrecurrence for many years or forever," he said. "That's hard to prove, but I think that should be the principle."

Viable structure may remain after excision of large tumors from the ear. But ocular tumors pose a wide range of problems, particularly if the tumor extends into the extraocular muscles.

If at all possible, surgeons should not sacrifice the important structures of the face or the facial nerve, he said.

A careful and early diagnosis of neurofibroma is important and can help distinguish NF1 from neurofibromatosis 2 (NF2), which is rarer, occurring in 1 in 40,000 births.

NF2 is characterized by multiple tumors on the cranial and spinal nerves and by other lesions of the brain and spinal cord. Its primary characteristic is bilateral tumors of the eighth cranial nerve, resulting in hearing loss beginning in the patient's teens or early 20s.

Because patients with neurofibroma may have recurrence throughout life and/or develop neurosarcoma, they need continued surgical surveillance, Dr. McKinnon said.

The National Institutes of Health is launching a study to investigate whether genetic differences explain differences in disease severity among patients with NF1. Participants must be at least 16 years old. For more information, write to jsloan@mail.nih.gov. ■

Distinguish Diaper Psoriasis From Seborrheic Dermatitis

BY SHARON WORCESTER
Southeast Bureau

BAL HARBOUR, FLA. — Diaper-area psoriasis, also known as napkin psoriasis, is a fairly common finding in children, Amy Paller, M.D., said at the annual Masters of Pediatrics conference sponsored by the University of Miami.

Although inclusion of this "rash" in the realm of psoriasis has been controversial, Dr. Paller said she "truly believes" it should be considered a psoriatic condition.

About 13% of children present with napkin psoriasis with dissemination; about 4% present with localized disease. The frequency of this condition in infants reflects the Koebner phenomenon—a response to the constant trauma to the skin that is a consequence of exposure to stool and urine.

It can be difficult to distinguish napkin psoriasis from seborrheic dermatitis, but there are certain characteristics that can help, said Dr. Paller, professor and chair of dermatology at Northwestern University, Chicago.

Plaques associated with napkin psoriasis are sharply defined, brightly erythematous, and larger than those in seborrheic dermatitis. They also tend to have drier scale, which in the diaper area often can be seen only when the skin is scratched.

This is caused by the constant moisture in the area, which also can obscure diagnosis in some cases, Dr. Paller noted.

For treatment, she uses a short course (about 3 weeks) of nonhalogenated topical steroid or topical calcineurin inhibitor and a lot of protective paste. ■