Childhood Dermatomyositis Is Almost Never Associated With a Cancer Risk

BY SHARON WORCESTER
Southeast Bureau

Destin, Fla. — Dermatomyositis is by far the most common form of idiopathic myositis in children, accounting for nearly 90% of cases, Dr. Brian Feldman said at the annual Rheumatology on the Beach. This differs from adult disease in that only about 14% of adult patients with myositis have dermatomyositis, with 11%-14% experiencing cancer-associated myositis, about 25% having overlap disease, and close to 50% having either polyomyositis or inclusion body myositis.

Dermatomyositis almost never occurs in children, thus a work-up for underlying malignancy is not necessary in this patient population, said Dr. Feldman, a professor at the University of Toronto.

The disease in children can range from mild to severe, and a papulovesicular heliotrope rash, usually on the extremities or trunk, is typical. Such a rash that occurs over the knuckles is known as Gottron's rash, and this rash is almost never present in children younger than 12 months, he said.

Another finding is periungual erythema resulting from capillary nailfold abnormalities (hazard ratio 0.5) and nail fold abnormalities (hazard ratio 0.6), and at 12 months, a combination of Gottron’s rash and weakness strongly predicted a chronic disease course, said Dr. Feldman, professor at the University of Toronto.

Gastrointestinal symptoms at diagnosis predicted significantly shorter time to remission (34 months vs. 84 months). The reason for this remains unclear, but in a poster on this data presented at the annual meeting of the American College of Rheumatology in November, Dr. Feldman and his colleagues noted that specific treatment regimens used early in the course of the disease may have contributed to the findings, which will be analyzed further.

The study also showed 60% of patients had chronic disease, 37% had monocyclic disease, and 3% had polyyclic disease, suggesting in children, remission is permanent. “When it’s gone, it’s almost always gone, and we can tell our patients that they are likely to stay in remission for the rest of their lives,” Dr. Feldman said.

Patients in the study had a mean age of nearly 8 years and a mean follow-up of nearly 6 years. The median time to remission was 4 years. About half remained within 4% years, and at 10 years, about one-third still had chronic active disease, he said.

—Sharon Worcester

Just to Be Sure: Overtreat in Suspected Kawasaki Disease

By NANCY WALSH
New York Bureau

New York — Don’t feel guilty about overdiagnosing and overtreating Kawasaki disease, Dr. Jeffrey R. Starke said at a meeting sponsored by the American College of Emergency Physicians.

“If you look at the risk-benefit ratio of treatment versus the complications if we don’t treat, it’s clear we should be on the side of overtreatment, especially in children younger than 12 months who are at high risk for developing severe coronary artery abnormalities,” Dr. Starke said.

These youngest patients also are more likely to present with incomplete Kawasaki disease. This diagnosis should be considered in infants under 6 months with fever for longer than 6 days and unexplained systemic inflammation. An incomplete Kawasaki disease diagnosis should also be considered in children with fever for over 5 days and two or three, rather than four, of the features of Kawasaki disease (see box).

In such a patient, if the C-reactive protein is 3 mg/dl or greater and/or the erythrocyte sedimentation rate is 40 mm/hour or more, supplemental laboratory criteria should be obtained. If three or more are present, the patient should have an echocardiogram and treatment should begin.

If there are fewer than three of the laboratory criteria, an echocardiogram is needed. If the echo shows cardiac abnormalities, treatment should begin.

Some continue the high dose for 14 days and then reduce the dose to 3.5 mg/kg per day, while others maintain the high dose only until 24-48 hours after the patient defervesces, and then switch to the low dose for 2 months for antplatelet effects.

“I don’t know of a single infectious disease expert or rheumatologist who doesn’t still use aspirin in addition to IVIG in the treatment of Kawasaki disease,” Dr. Starke said.

Some data suggest some benefit of adding corticosteroids to treatment. In a recent prospective randomized trial of 178 Kawasaki patients, children on prednisolone with IVIG had a shorter duration of fever and a faster fall in C-reactive protein, but no difference in coronary artery dilation at 1 month (J. Pediatr. 2006;149:336-41).

If the fever persists or returns more than 36 hours after completion of the IVIG infusion, a second dose can be given.

“Just be sure: It’s very common for kids to continue to run a fever the day after you give the IVIG dose, even as other symptoms are improving,” he said.

Clinical Diagnostic Features of Kawasaki Disease

**Diagnostic Criteria**

Fever greater than 39 degrees C for 5 days, plus four of the following:

- Conjunctivitis, usually bulbar and bilateral
- Mucovascular changes such as redness and cracking
- Rash, either generalized or local
- Enlarged cervical lymph nodes, usually unilateral and tender
- Peripheral changes such as swelling or peeling

**Incomplete Kawasaki Supplemental Laboratory Criteria**

- Albumin 3 g/dl or less
- Anemia for age
- Elevation of alanine aminotransferase
- Platelets 450,000/mm3 or more after 7 days
- White blood cell count 15,000/mm3 or higher
- Urine white blood cell count 10/HPF or more

Sources: Dr. Starke; Circulation 2004;110:2471-71.