Skin Problems May Provide Clues to GI Disease

BY DOUG BRUNK
San Diego Bureau

LA JOLLA, CALIF. — Certain skin conditions may provide clues to the diagnosis of underlying gastrointestinal disease in children, ranging from epithelial defects, polyposis, or vascular syndromes to autoimmune and allergic disease.

The latter may involve overlapping issues between the skin and the GI tract,“ Dr. Magdalene A. Dohil said at a meeting sponsored by Rady Children’s Hospital and the American Academy of Pediatrics.

Diseases of the GI tract that commonly involve some form of cutaneous manifestation include:

- **Epidermolysis bullosa**: This condition presents with different degrees of skin fragility and blister formation. The severity “really depends on the underlying molecular defect,” said Dr. Dohil, who is a pediatric dermatologist at Rady Children’s Hospital, San Diego. “GI disease in epidermolysis bullosa is extremely common, particularly in the recessive dystrophic type,” (in which) almost 80% of children are affected by (dysphagia).

- Other symptoms may include lingual adhesions and microstomia; esophageal disease including strictures, webs, and antral atony, and pseudodiverticula leading to feeding problems and ultimately protein-energy malnutrition, anemia; and vitamin and mineral deficiency.

- **Blue-rubber bleb nevus syndrome (BRBNS)**. This disease causes multifocal venous malformations in the skin and GI tract. Most cases are sporadic, and histology demonstrates intact epithelium but insufficient smooth muscle. Dr. Dohil described the case of a child who presented with venous malformations on the bottom of the foot, which resembled common warts at first glance. “But when you palpate [them], they are soft and compressible.”

- **Common complications of BRBNS include bleeding, chronic anemia, and the need for blood transfusions.** Treatment often involves different degrees of surgical intervention including wedge resection, polypectomy, stent ligation, band ligation, and sometimes bowel resection. “Medical treatment attempts because they have been very successful are not because these are pro liferative tumors, so we don’t expect them to respond to corticosteroids or interferon,” said Dr. Dohil, also of the University of California, San Diego. Capsule endoscopy “facilitates the diagnosis and follow-up of children who need endoscopic intervention and assessment.”

- **Peutz-Jeghers syndrome**: “Autosomal dominant type,” Dr. Dohil said, and “high risk of developing invasive carcinoma.” In fact, their cumulative risk of developing cancer is 93%, most commonly cancers of the breast, colon, and pancreas, noted Dr. Dohil.

- **Cowden’s disease**: This condition, known as multiple hamartoma-neoplasia syndrome, causes hamartomas that involve the skin, intestine, breast, and thyroid.

- **Henoch-Schönlein purpura**: “Medical treatment attempts haven’t been very successful,” Dr. Dohil said. Such a blunted GI tract doesn’t bode well for the absorptive functions that it’s intended for.

Dermatitis herpetiformis (Duhring’s disease) is considered a cutaneous manifestation of celiac disease. This condition affects about 25% of celiac disease patients and is marked by a pruritic eruption of lesions that may be symmetrical, erythematous, papular, vesicular, or bullous. It commonly occurs in the trunk area and on the back of the forearm and elbow. These lesions “are fairly uncommon in children, and when they do occur they may not be very distinct,” she said. Recently a variety of skin conditions such as seborrheic, urticaria, vitiligo, and alopecia areatae have been linked to celiac disease. However, since they are fairly non-specific, skin biopsies with direct immunofluorescence and antibody studies of gladin, endomysium, and transglutaminase are often needed to confirm the diagnosis. Treatment includes dapsone and a gluten-free diet.

Dr. Dohil reported that she had no relevant disclosures to make.

New Findings on Chronic Urticaria Refine Screening, Improve Outcomes

BY SHARON WORCESTER
Southeast Bureau

SAN ANTONIO — Treatment of chronic urticaria can be challenging, but recent findings on the condition may help improve outcomes, Dr. Aniko Kobza Black reported at the annual meeting of the American Academy of Dermatology.

One new advance is the usefulness of the autologous serum skin test (ASST) in screening for autoimmune urticaria, even if patient history, and ASST status before try-out are not adequate, it is important to re-assess disease severity, patient history, and ASST status before trying the second- and third-line treatments. There is no risk, Dr. Black reported, of the three types of treatments can be combined, however, “and indeed they usually have to be,” Dr. Black said.

The role of *Helicobacter pylori* in chronic urticaria is controversial. About 40% of patients have abdominal symptoms, but no evidence shows *H. pylori* infection causes the condition. It may be that it plays an indirect role in genetically predisposed individuals, but this remains unclear.

First-line treatment for chronic urticaria remains low-dose antihistamines. The use of doses above recommended levels remains controversial—it seems to be clinically effective in some patients, but no trials have shown efficacy with the approach.

In rare cases, antihistamines may actually aggravate urticaria. Allergic reactions can occur in minutes but usually occur after 6 hours, and they have been seen with each type of antihistamin. No definite cause is known, but the reactions may be the result of a direct effect on mast cells.

When deciding on treatment, first consider side effects, then ease of administration, and then cost, she advised. It is not possible to predict which treatments will be effective in a given patient. When first-line antihistamine treatments and combinations are inadequate, it is important to re-assess disease severity, patient history, and ASST status before trying the second- and third-line treatments. There is no risk, Dr. Black said, of the three types of treatments can be combined, however, “and indeed they usually have to be,” Dr. Black said.

The patient cohort, accumulated over 129 international sites, was divided into two arms, with 547 patients receiving ceftriaxone and 281 receiving the glycopeptide antibiotic vancomycin plus the third-generation cephalosporin cefazolin. Respectively, the two study arms consisted of 63% and 64% men and had mean ages of 53 and 52 years. The proportion of patients completing the trial was 92% in the ceftriaxone arm and 90% in the comparator arm. Patients in the ceftriaxone arm received 500 mg for 120 minutes every 8 hours. In the comparator group, the starting dose was 1,000 mg vancomycin infused over 60 minutes every 12 hours plus 1,000 mg cefazolin infused over 120 minutes every 8 hours. The mean duration of treatment in the clinically evaluable population was about 28 days in both arms, the authors noted.

At the test-of-cure (TOC) visit (after 6-17 days of treatment) for the evaluable patients, clinically evaluable 485 (91%) ceftriaxone-treated patients and for 220 of 244 (90%) comparator-treated patients, the researchers noted.