SECTION EDITOR: ENID GILBERT-BARNESS, MD

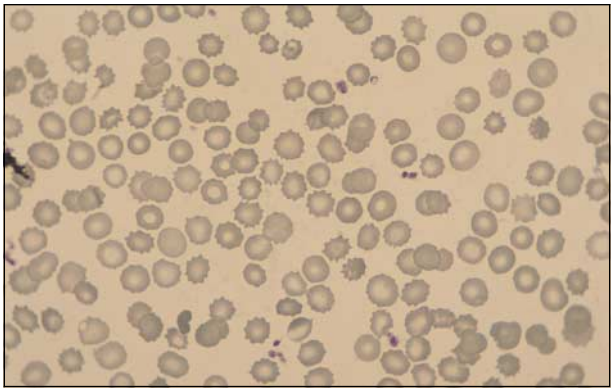
Pathological Case of the Month

Ahmet Karadag, MD

A 7-MONTH-OLD BOY with malnutrition and chronic diarrhea came to the emergency department with dehydration and metabolic acidosis. His medical history revealed that he had diarrhea, vomiting, and abdominal distension for 1½ months. His stool was defined as foul-smelling, greasy, and bulky. He was born from nonconsanguineous parents following an uneventful pregnancy. He had been breastfed until he was 3 months old, at which time he began receiving formula, rice, and biscuits.

From the Department of Pediatrics, Fatih University Medical School, Emek Ankara, Turkey.

At physical examination, his weight was 5800 g (<3rd percentile), his height was 68 cm (<75th percentile), and his head circumference was 42 cm(<10th percentile). He was dehydrated and his abdomen was severely distended but there was no organomegaly. Results of laboratory investigations were compatible with metabolic acidosis and mild hypokalemia. A complete blood cell count showed hemoglobin to be 11.4 g/dL; hematocrit, 35.3%; mean corpuscular volume, 71.9 fL; white blood cell count, $9.2 \times 10^3/\mu L$ ($9.2 \times 10^9/L$); and platelets, $220 \times 10^3/\mu L$ ($220 \times 10^9/L$). A peripheral blood smear revealed 36% granulocytes, 52% lymphocytes, 10% monocytes, 2% bands, and generalized acanthocytosis was noticed, involving more than 80% of erythrocytes (**Figure**).



©2002 American Medical Association. All rights reserved.
Downloaded From: http://archpedi.jamanetwork.com/ on 12/02/2016

Diagnosis and Discussion

Celiac Disease with Acanthocytosis

Figure. Acanthocytosis is seen on the blood smear.

Biochemical tests of plasma for renal and liver functions were considered normal. Results of microscopic examination of feces was normal except for increased lipid content, and microbiologic test results were also negative. Antibody titers against toxoplasmosis, cytomegalovirus, herpes simplex virus types 1 and 2, and rubella were not suggestive. Serum lipids were found as follows: total cholesterol, 57 mg/dL (1.47 mmol/L); high-density lipoprotein, 31.3 mg/dL; low-density lipoprotein, 7.3 mg/dL; and very low-density lipoprotein, 18.4 mg/dL. Lipoprotein electrophoresis was found in normal limits; consequently, abetalipoproteinemia was not considered. Other disorders that cause malabsorption and failure to thrive, such as cystic fibrosis, were ruled out. Blood and urine amino acid concentrations were normal. Antigliadin immunoglobulin (Ig) A and IgG antibody titers were 58.4 (normal, 0-22) U/mL and 50.5 (normal, 0-27) U/mL, respectively. Endomysium IgA antibody was positive, which was highly suggestive for celiac disease (CD).

He began receiving a gluten-free diet and subsequently significant improvement of his clinical situation was observed. He gained weight dramatically and diarrhea ceased. Two weeks later, acanthocytosis was not seen on his blood smear.

Celiac disease, also called gluten-sensitive enteropathy, is a permanent intestinal intolerance to dietary wheat gliadin and related proteins that produces lesions in genetically susceptible individuals.[1] Clinical features differ depending on the age of onset. In children, the onset of the disease is within the first through third years of life after exposure to gluten, and they exhibit a classic syndrome of chronic diarrhea, failure to thrive, and abdominal distention.[2] Different neurologic metabolic symptoms, such as epilepsy with cerebral calcification, ataxia, and intellectual deterioration, can occasionally accompany CD.[2] Alopecia is another extraordinary finding that has been reported in children with CD.[3] Acanthocytes are dense, contracted red blood cells with several irregularly spaced thorny projections on the surface.[4] Acanthocytosis is seen in different kinds of disorders. Although the exact mechanism of formation of acanthocytes is not clear, reversal of the usual phosphatidylcholine-sphingomyelin ratio is considered to be a possible mechanism in certain diseases, such as abetalipoproteinemia.[4]

To the best of our knowledge, this is the first report of the coexistence of acanthocytosis and CD in children. We do not think that this finding indicates an association because the acanthocytosis was reversed after prompt treatment. We suggest that CD, which is a more common cause of malabsorption in children than abetalipoproteinemia, should be considered when acanthocytosis is seen on a peripheral blood smear.

Accepted for publication April 18, 2000.

Corresponding author: Ahmet Karadag, MD, Department of Pediatrics, Fatih University Medical School, Ciftlik Caddesi No. 57 06510, Emek Ankara, Turkey (e-mail: kara_dag@hotmail.com).

REFERENCES

1. Troncone R, Greco L, Auricchio S. Gluten-sensitive enteropathy. *Pediatr Clin North Am.* 1996;43:355-373.
2. Nehra V. New clinical issues in celiac disease. *Gastroenterol Clin.* 1998;27:453-464.
3. Naveh Y, Rosenthal E, Ben-Arieh Y, Etzioni A. Celiac disease associated alopecia in childhood. *J Pediatr.* 1999;134:362-364.
4. Gladder EB, Lukens JN. Acanthocytic disorders. In: Lee GL, Foerster J, Lukens J, Poraskevas F, Greer JP, Rodgers GM, eds. *Wintrobe's Clinical Hematology.* Baltimore, Md: Williams & Wilkins; 1999:1150-1151.

Submissions

The Editor is seeking submissions for a new feature, *Clinical Problem Solving*, which will combine *Picture of the Month*, *Radiological Case of the Month*, and *Pathological Case of the Month*. Our aim is to demonstrate the thinking process of a master clinician involved in approaching a patient with an unknown disease. The discussion of such cases should place the clinician's expertise into the context of the prevailing medical literature on the topic. Manuscripts should be between 3000 and 4000 words and may include photographs and radiographs.

©2002 American Medical Association. All rights reserved.
Downloaded From: http://archpedi.jamanetwork.com/ on 12/02/2016

6. Allen KF, Moss AJ, Giovino GA, Pierce JP. *Teenage Tobacco Use: Data Estimates From the Teenage Attitudes and Practices Survey: United States, 1989*. Advanced data 224. Hyattsville, Md: National Center for Health Statistics; 1992
7. Bal DG, Kizer KW, Felten PG, Mozar HN, Niemeyer D. Reducing tobacco consumption in California: development of a statewide anti-tobacco use campaign. *JAMA*. 1990;264:1570-1574.
8. Sussman S, Lichtman K, Ritt A, Pallonen UE. Effects of thirty-four adolescent tobacco use cessation and prevention trials on regular users of tobacco products. *Substance Use Misuse*. 1999;34:1469-1503.

Preventing Unnecessary Emergency Department Visits for "Albuterol Nebs"

The article by Tien et al[1] reminds us that delivering albuterol with a metered-dose inhaler with a valved spacer (MDI) for acute asthma attacks has several advantages over using a nebulizer. They carefully document equivalent or improved efficacy, more rapid delivery, and lower cost. I would add that using an MDI in the emergency department (ED) provides an opportunity to reteach the parent how to properly administer bronchodilators by MDI at home. That may eliminate the need for unnecessary return visits to the ED for another "neb bail-out." The good advice in this article also applies to clinic and office treatment of asthma attacks.

Barton D. Schmitt, MD
Child Health Clinic
The Children's Hospital
1056 E 19th Ave, Box 085
Denver, CO 80218

1. Tien I, Dorfman D, Kastner B, Bauchner H. Metered-dose inhaler: the emergency department orphan. *Arch Pediatr Adolesc Med*. 2001;155:1335-1339.

In reply

We thank Dr Schmitt for his comments and certainly agree that the use of MDIs for acute asthma attacks in the ED would present an opportunity to educate parents and patients about the proper use. It also would allow for reinforcement of the benefits of MDI use at home, including improved portability, lower cost, and reductions in the total time required to administer treatment. There continue to be, however, barriers to translating this knowledge into a change in physician behavior. As we have previously described, medical decisions in which the physician has more discretion are often greatly influenced by the physician's perception of patient preference.[1] This study demonstrated that 24% of pediatric ED physicians are under the impression that patients and parents prefer nebulizers over MDIs when they come to the ED for acute asthma. However, in studies comparing patient and parent satisfaction with MDIs vs nebulizers for the treatment of acute asthma in the ED, MDIs were preferred.[2,3] Until the medical community embraces the benefits of MDI use for acute asthma in children, parents will continue to receive inconsistent messages about their use in the home.

Irene Tien, MD
Howard Bauchner, MD
Department of Pediatics
Boston Medical Center
1 Boston Medical Center Pl, MAT 6
Boston, MA 02118

1. Bauchner H, Simpson L, Chessare J. Changing physician behavior. *Arch Dis Child*. 200;84:459-462.
2. Schuh S, Johnson DW, Stephens D, Callahan S, Winders P, Canny GJ. Comparison of albuterol delivered by MDI with spacer vs a nebulizer in children with mild acute asthma. *J Pediatr*. 1999;135:22-27.
3. Fugelsang G, Pedersen S. Comparison of nebuhaler and nebulizer treatment of acute severe asthma in children. *Eur J Respir Dis*. 1986;69:109-113.

Correction

Error in Byline. In the special feature titled "Pathological Case of the Month: Celiac Disease With Acanthocytosis," published in the March issue of the ARCHIVES (2002;156:291-292), on page 291, the byline should have read, "Ahmet Karadağ, MD; E. Esra Önal, MD; Füsun K. Uysal, MD; İ. Safa Kaya, MD; Uğur Dilmen, MD."

©2002 American Medical Association. All rights reserved.
Downloaded From: http://archpedi.jamanetwork.com/ on 12/02/2016