Case Challenge

A 3-Year-Old Girl with Eye Pain

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A 3-year-old girl presents with bilateral eye pain. The history of the complaint began 3 weeks earlier with headache, photophobia, and intermittent eye pain. During this time, no erythema or discharge from the eyes is noted and no other symptoms reported.

Her medical history is positive for multiple parental-diagnosed allergies and chemical sensitivities, resulting in behavioral problems, frequent loose stools, and abdominal pain. Her parents express concern that she may have gastroesophageal reflux disease and irritable bowel syndrome.

Examination reveals an alert but irritable 3-year-old girl with normal vital signs. Her development is delayed and behavior is abnormal, with extreme fear and anxiety. Abnormal findings related to her eyes include dry, lusterless corneas, visible ulcerations, and a large area of hyperkeratosis with some erythema of the right eye. She also has rough, dry skin. Under anesthesia, an ophthalmologic examination determines that the corneal ulcer in the right eye requires an emergent corneal transplant.

For diagnosis, see page 360

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at Pediatrics@Healio.com.
A detailed review of the patient’s dietary history reveals a severely restricted diet that is limited to chickpea milk, bison meat, and salicylate-free vegetables. The patient’s 10-year-old sister was placed on a similar diet at age 1 year that resulted in scurvy and rickets by age 3 years. These findings are suspicious for possible vitamin-A deficiency, and the patient is admitted to the hospital for further evaluation and treatment. The patient’s severely restricted diet has resulted in micronutrient malnutrition. Laboratory results show hemoglobin of 7.5 g/dL, iron of 35 mcg/dL, transferrin saturation of 12%, prealbumin of 6 mg/dL, and vitamins A and C below assay limits. The patient exhibits profound oral aversion as well as behavioral difficulties as a result of her long-standing dietary restrictions, which proved very problematic as an appropriate diet was reintroduced.

**Diagnosis:**

Vitamin-A Deficiency

This previously healthy, unimmunized child has severe vitamin-A deficiency with xerophthalmia, corneal ulcerations, follicular hyperkeratosis, scurvy, anemia, and malnutrition. During the hospitalization, the parents requested strict adherence to a “chemical-free” environment and avoidance of medications with preservatives and artificial coloring. However, upon further investigation, the mother, a health care provider, was unable to provide any supporting evidence for the chemical sensitivities; in fact, the patient was previously seen by more than a dozen providers without substantiation of these diagnoses. Against the parents’ wishes, vitamin-A supplementation was provided to correct the deficiency and decrease her risk of serious infections from damaged skin, mucous membrane barriers, and decreased cellular immunity. Immunizations were provided, particularly the measles, mumps, rubella (MMR) vaccine, because of the significant mortality risk associated with serious measles infection and vitamin-A deficiency. After restricting parental visits, the patient’s behavior gradually began to normalize, and she was ultimately discharged to the custody of child protective services.

**DISCUSSION**

Vitamin A is vital for maintaining the integrity of epithelial function, in particular the mucous membranes of the eye, respiratory, urinary, and intestinal tracts, which are essential barriers to pathogens. The most clinically apparent signs of vitamin-A deficiency are night blindness and xerophthalmia, which is the development of dry, scaly layers of the cornea. Ultimately, the abnormally keratinized cornea develops plaques, referred to as Bitot spots (see Figure 1, page 359). Additional findings include dry, hyperkeratotic areas of skin, particularly of the arms and legs (see Figure 2).

Normal adult hepatic stores of vitamin A may take more than 1 year to be depleted in the absence of any dietary source. The neonate, however, has little hepatic reserve, and many growing children only have a few weeks of vitamin A reserve, which may be rapidly exhausted during periods of acute illness or stress. Infant formulas are fortified with vitamin A to prevent such deficiency during the first year of life. Infants are particularly at risk from nutritionally inadequate diets because of the high nutritional needs associated with their rapid growth rate. Those who are breast-fed may be protected, but if weaned to deficient diets they may become seriously malnourished within weeks.

Vitamin-A deficiency is one of the world’s top five major malnutrition...
problems, estimated to affect more than 124 million children worldwide.\(^1\) Vitamin-A deficiency is known to occur in developing countries, most commonly in children younger than age 5 years. The association of xerophthalmia with poor nutrition and low socioeconomic status was identified in the early 1800s;\(^2\) thus, even before the discovery of vitamin A and the elucidation of its biochemical mechanisms, the ocular manifestations of this deficiency and treatment were understood. Despite recognizing this condition more than 200 hundred years ago, xerophthalmia remains a major public health problem in developing countries and is an area of active research. The burdens of childhood blindness and mortality associated with vitamin A deficiency are especially substantial in the underdeveloped countries of Asia, Africa, and Latin America. Vitamin A deficiency has long been identified as the leading cause of pediatric blindness in the world.

Ongoing interest in fad diets by parents, and consequent development of nutritional problems in their children, have been well described in children who are fed restricted diets. The parents’ opinion about certain foods, their strong likes or dislikes, their willingness to try new foods, and their eating patterns greatly influence the child’s dietary intake. Their influence on food patterns is critical in the development of food preferences. Elimination diets (such as the Kaiser-Permanente diet developed by Benjamin Feingold, MD\(^3\)) that focus on the elimination of artificial food colors and salicylate-containing foods have garnered significant attention in hopes they might offer symptom improvement in children with attention-deficit/hyperactivity disorder. The patient presented here illustrates the ensuing nutritional complications that can stem from a strong parental belief in a restricted diet.

**CONCLUSION**

Subclinical vitamin-A deficiency is a worldwide health problem in young children. The World Health Organization estimates that 70 million to 80 million children have subclinical vitamin A deficiency.\(^1\) The problem is most likely under-recognized in the United States and other developed countries, who do not normally consider their citizens to be malnourished. Lack of a simple screening test to measure subclinical deficiency contributes to this problem. Extreme dietary preferences may complicate the nutritional status assessment in our pediatric population. Consequently, focused dietary assessment of micronutrient intake is a necessary component of health care maintenance, especially for toddlers and preschool-aged children. Dietary counseling and vitamin supplementation for high-risk children is a necessary health promotion intervention. When a dietary deficiency exists and parents reject treatment, notification of local child protective services is indicated.

**REFERENCES**

3. Feingold BF. *Why Your Child is Hyperactive*. New York: Random House; 1975,