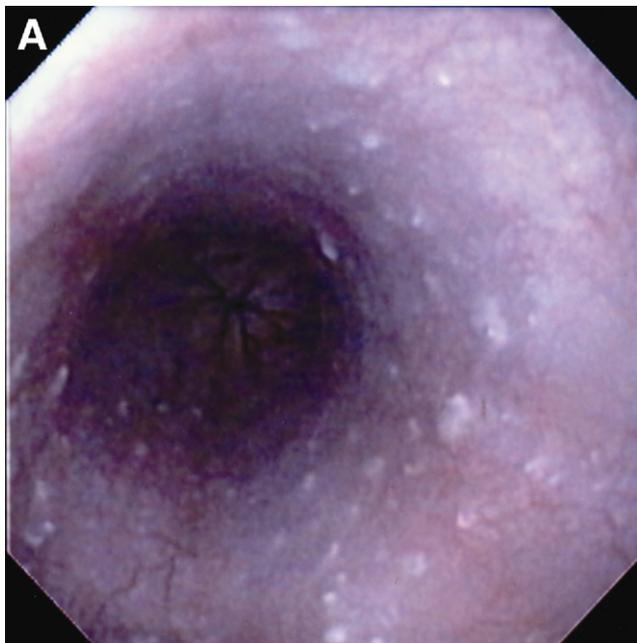


Electronic Clinical Challenges and Images in GI

David A. Katzka and David L. Jaffe, Section Editors

An Uncommon Cause of Esophagitis



Question: A 9-year-old boy with Landau-Kleffner syndrome (acquired aphasia and convulsive disorder) was transferred for evaluation of melanotic stools and worsening anemia. He had been admitted to an outside hospital 1 week before transfer because of vomiting, worsening oral intake, lethargy, and dehydration resulting from a urinary tract infection. During the hospitalization he developed melena with associated decrease in hemo-

globin from 11.1 to 7.7 g/dL, prompting his transfer. Other significant medical history included malnutrition (attributed to a diet consisting exclusively of milk, pretzels, and chicken nuggets), seizures (controlled with oxcarbazepine and levetiracetam), and progressive blindness (recently attributed to vitamin A deficiency). Examination was significant for a body mass index <3rd percentile, pallor, tachycardia, and occult blood positive stool. Upper endoscopy, done to evaluate for a bleeding source, revealed shallow antral erosions without active bleeding. A rapid urease test was negative. The esophageal mucosa was pale, with furrowing along its entire length; multiple pin-point, white, papules were seen (Figure A). What is the etiology of the esophageal findings?

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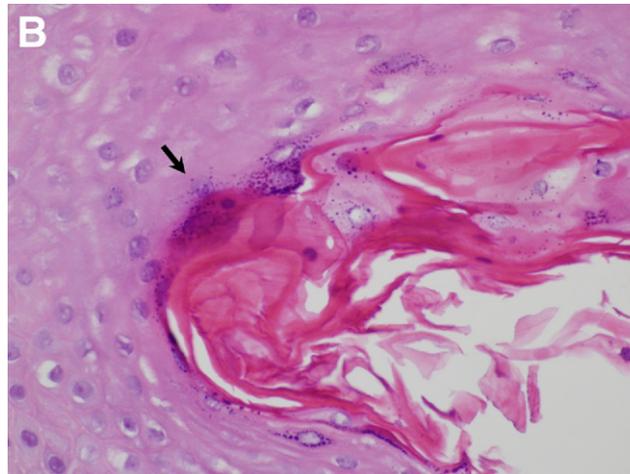
Conflicts of interest

The authors disclose no conflicts.

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Answer to the Clinical Challenges and Images in GI Question: Image 1: Esophageal Hyperkeratosis Secondary to Vitamin A Deficiency

Histologic evaluation showed hyperkeratosis and mild acute esophagitis (Figure B). A diagnosis of esophageal hyperkeratosis secondary to vitamin A deficiency was made. The differential diagnosis of the endoscopic features of esophageal hyperkeratosis includes squamous papilloma, human papilloma virus (HPV)-induced papilloma, and eosinophilic esophagitis. Esophageal biopsy is invaluable in securing a diagnosis. Hyperkeratosis is characterized by a dark eosinophilic acellular layer of keratin overlying a thickened stratified squamous epithelium. Beneath the keratin lies a granular layer containing cells with intensely basophilic-stained cytoplasmic granules known as keratohyalin granules (*arrow*).

Esophageal hyperkeratosis has been seen in association with aberrations of fat-soluble vitamins, vitamin A deficiency, and vitamin E excess, suggesting an etiologic role.¹ Esophageal involvement may complicate other hyperkeratotic states, such as Darier's disease and tylosis. Esophageal squamous cell carcinoma has been associated with hyperkeratotic states of differing etiologies, including Darier's disease, tylosis, and HPV-induced papillomatosis.² The risk of esophageal carcinoma in HPV papillomatosis differs among the various HPV types: high-risk types include HPV 6, 16, and 18.² Similarly, the role of HPV in esophageal carcinoma varies by geographic location; a high association is found in Asia and South Africa, whereas a low association is seen in Western Europe.² Interestingly, similar to other hyperkeratotic states, vitamin A deficiency has also been associated with esophageal carcinoma. In 1 study, vitamin A deficiency was found in 88% of adults with esophageal squamous cell carcinoma.³ Mucosal changes owing to vitamin A deficiency, including hyperkeratosis, improve after correction of the deficiency with supplementation. Repeat endoscopy 4 months after initiation of vitamin replacement resulted in endoscopic and histologic improvement.

References

1. Mak KM, Leo MA, Lieber CS. Effect of ethanol and vitamin A deficiency on epithelial cell proliferation and structure in the rat esophagus. *Gastroenterology* 1987;93:362-370.
2. Lavergne D, de Villiers EM. Papillomavirus in esophageal papillomas and carcinomas. *Int J Cancer* 1999;80:681-684.
3. Mellow MH, Layne EA, Lipman TO, et al. Plasma zinc and vitamin A in human squamous carcinoma of the esophagus. *Cancer* 1983;51:1615-1620.

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