

Esophageal Intramural Pseudodiverticulosis With Tracking in a Child With Autosomal Dominant Hyper-IgE Syndrome



FIGURE 1. Barium esophagogram with intramural contrast seen running parallel with lumen of esophagus.

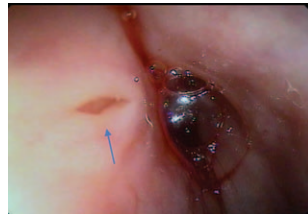


FIGURE 2. Single diverticulum seen on esophagogastroduodenoscopy (EGD) (arrow).

An 11-year-old boy from Saudi Arabia with autosomal dominant hyper-IgE syndrome presented after an incidental finding of esophageal pneumotosis on chest computed tomography. Barium esophagogram demonstrated intramural tracking of contrast from the mid to distal esophagus suggestive of esophageal intramural pseudodiverticulosis (EIP) with tracking (Fig. 1). Esophagogastroduodenoscopy (EGD) confirmed the presence of a single diverticulum (Fig. 2), and culture of esophageal biopsies grew *Saccharomyces cerevisiae* and *Candida glabrata*. Esophageal mucosa showed marked eosinophilia and lymphocytosis, with basal cell hyperplasia. The patient returned home before tailored therapy could be initiated.

EIP is a condition in which the esophageal submucosal excretory glands form outpouchings, thought to be secondary to obstruction or dilatation from inflammation or altered motility (1). Findings include flask-shaped outpouchings on barium esophagogram, and openings of pseudodiverticula seen on EGD (1,2). Tract

formation is rare, and typically indicates more diffuse EIP affecting more of the esophagus (1). Although this patient was without gastrointestinal symptoms, EIP most commonly presents as dysphagia, and carries the risk of perforation and mediastinitis (3,4). In this case, we hypothesize that the presence of infection fostered the formation of EIP.

This is the first reported case of EIP in hyper-IgE syndrome and highlights the diagnostic utility of EGD paired with an unusual radiographic finding.

Submitted by:

*Frances C. Lee, [†]Vignesh Ramachandran, [‡]Kevin P. Shah, [§]Filiz O. Seeborg, and ^{||}Douglas S. Fishman

*Department of Pediatrics, [†]Department of Medical Education, [‡]Department of Immunology, Allergy, and Rheumatology, Baylor College of Medicine, [§]Texas Children's Hospital, and ^{||}Section of Pediatric Gastroenterology, Hepatology, and Nutrition, Baylor College of Medicine, Houston, TX.

Address correspondence and reprint requests to Frances C. Lee, MD, Department of Pediatrics, Baylor College of Medicine, One Baylor Plaza BCM 320, Houston, TX 77030 (e-mail: frances.lee@bcm.edu).

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Colonic Diverticulitis in an Infant With Hemophagocytic Histiocytosis and Prolonged Glucocorticoid Exposure

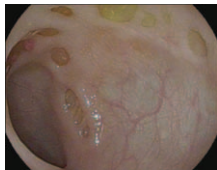


FIGURE 1. Diverticula.



FIGURE 2. Friable diverticula.

A 9-month-old boy with hemophagocytic histiocytosis (HLH) diagnosed at 4 months of age and treated since then with high-dose oral dexamethasone and Emapalumab presented with hematochezia associated with a hemoglobin drop from 10.5 to 8.8 g/dL for 2 days. Vital signs were normal. Exam was remarkable for Cushingoid appearance and normal perianal and rectal exam. Blood and stool studies for infection and Meckel diverticulum scan with H2 blockade were negative. Subsequent colonoscopy with a PENTAX EG-2770K endoscope demonstrated numerous (>10) diverticula in the rectosigmoid colon starting at 10 cm from the anal verge and extending 15 cm proximally (Fig. 1). Some had mild oozing, but none was actively bleeding (Fig. 2). Due to the concern for risk of perforation, the colonoscopy was aborted. He eventually underwent bone marrow transplant. He had no subsequent episodes of hematochezia and has not undergone repeat colonoscopy or computed tomography studies.

Colonic diverticula are extremely rare in children, although some genetic conditions including Ehlers-Danlos and Marfan syndrome have been linked to the development of diverticula (1–3). Gluco-

corticoids have been associated with diverticular disease in adults (4). An association between glucocorticoids or HLH with diverticular disease in children has not been reported.

Submitted by:

Liz Febo-Rodriguez, Seiji Kitagawa, and Eric Chiou

Pediatric Gastroenterology Department, Baylor College of Medicine, Houston, TX.

Address correspondence and reprint requests to Liz Febo-Rodriguez, MD, 6701 Fannin St, Suite 101000, Houston, TX 77030-2608. Address for reprints: Liz Febo-Rodriguez, MD, 5605 Newcastle St, Bellaire, TX 77401 (e-mail: liz.febo-rodriguez@bcm.edu, ljfebo@gmail.com)

L.F.-R. meets 4 criteria recommended by the ICMJE and evaluated patient and completed procedure; S.K. meets 4 criteria recommended by the ICMJE and evaluated patient and supervised procedure; and E.C. meets 4 criteria recommended by the ICMJE and evaluated patient and supervised procedure.

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The authors report no conflicts of interest.

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