

Guess the Case from the Ochsner Archives

David J. Licini, MD,* William S. Richardson, MD†

*Department of Orthopedics, Ochsner Clinic Foundation, New Orleans, LA

†Department of Surgery, Ochsner Clinic Foundation, New Orleans, LA

INTRODUCTION

A 29-year-old female with a past medical history of gastroesophageal reflux (GERD) presented to her primary care doctor with intractable nausea and vomiting 3 weeks after the cesarean delivery of a healthy baby. The patient stated that she first began feeling nauseated and vomited during the second half of her pregnancy. At that time, the patient was diagnosed with hyperemesis gravidarum. It was thought that her symptoms would resolve after the delivery. Since the birth of her child though, her nausea and vomiting had become worse. The symptoms occurred daily. She said that she had trouble swallowing solids and liquids, and anytime she tried to eat she would vomit it right back up. The patient did have significant weight loss from her inability to keep food down but denied any fevers/chills, hematemesis, recent blood transfusions, icterus, constipation/diarrhea, increased cough, or hoarseness. The patient was initially admitted to an outside hospital with the diagnosis of postoperative ileus.

The initial physical examination was unremarkable. The patient was afebrile, and vital signs were within normal limits. She was well-nourished and anicteric. The abdomen was non-distended and had only a well-healed low transverse abdominal incision. Bowel sounds were normoactive, the abdomen was soft with no epigastric or right upper quadrant pain, and hepatosplenomegaly was not present. Results of the laboratory tests were unremarkable.

Preoperative image from the barium swallow study is shown in Figure 1 and from the computed tomography (CT) scan in Figure 2.

Address correspondence to:
William S. Richardson, MD
Ochsner Medical Center
1514 Jefferson Highway
New Orleans, LA 70121
Tel: (504) 842-5874
Fax: (504) 842-5875
Email: dlicini@ochsner.org

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What is the diagnosis, and what treatment would you recommend?

WHAT IS THE DIAGNOSIS?

This patient has achalasia. This was confirmed with esophageal manometry, which showed an absence of coordinated esophageal peristalsis and an increased resting lower esophageal sphincter pressure with incomplete relaxation. The patient's barium swallow study shows severe dilation of the distal esophagus with a bird-beak deformity near the gastroesophageal (GE) junction. There is no contrast entering the stomach (Figure 1). The CT scan also shows a dilated esophagus with narrowing of the esophagus at the GE junction (Figure 2).

The patient underwent a laparoscopic Heller myotomy. She tolerated the procedure well, was started on a clear liquid diet, and discharged on postoperative day 1. The patient was advised to follow a full liquid diet for the first week, a soft mechanical diet for the second week, and a normal diet thereafter. The symptoms were resolved.

DISCUSSION

Achalasia is a term that was first used by Sir Thomas Willis in the 17th century. The Greek word literally means "failure to relax." The etiology of achalasia is unknown, but the underlying problem is degeneration of the ganglion cells in the myenteric plexus of the esophageal wall.¹ This degeneration leads to loss of peristalsis in the distal esophagus and failure of the lower esophageal sphincter (LES) to relax. Although both of these problems are present in the disease, the primary problem seems to be the defect in LES relaxation. Treatment is typically focused on this defect.

Achalasia is uncommon and affects an estimated 1 in 100 000 persons. Hyperemesis gravidarum is approximately 2000 times more common than achalasia.^{2,3} Differentiating between achalasia and other disorders is difficult because many disorders, positively GERD, can mimic achalasia clinically. Asking the right questions may be helpful, but often the diagnosis of achalasia is not entertained until further studies have been performed. Achalasia typically presents between the ages of 25 and 60 with a major symptom being dysphagia for both solids and liquids.⁴ The timing of these symptoms is important because dysphagia of liquids prior to

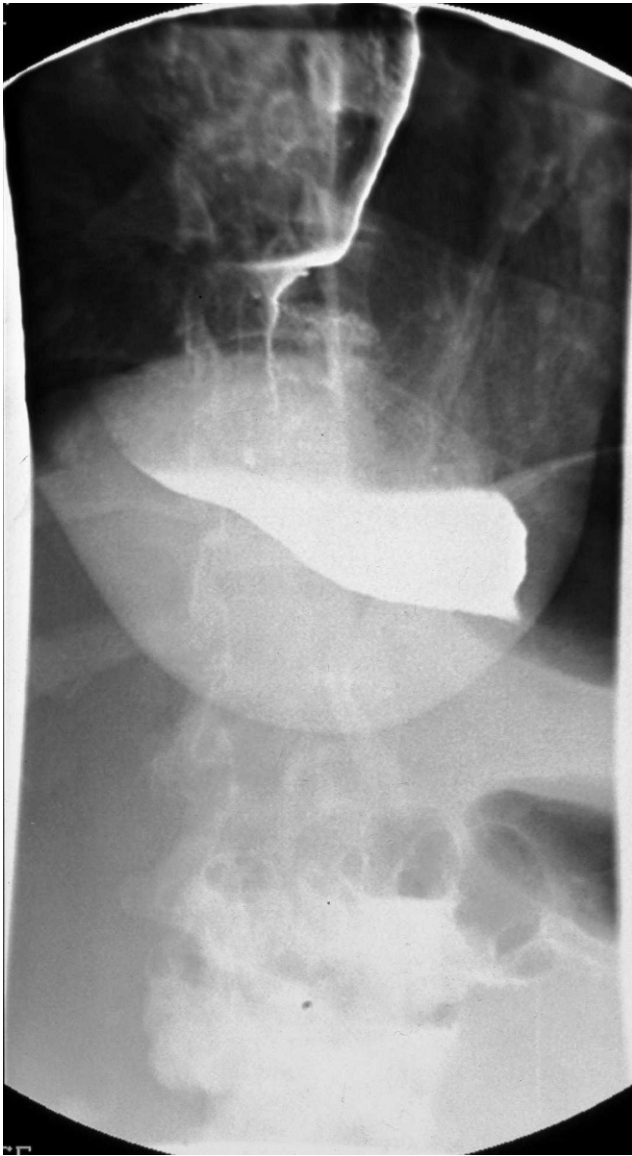


Figure 1. Representative image from the barium swallow study.

dysphagia of solids may represent pseudoachalasia rather than true achalasia. Another common symptom is difficulty belching, although most patients will not provide this information unless prompted. Regurgitation, chest pain, and weight loss are also common associated symptoms.

Making the diagnosis of achalasia is often difficult and may be delayed for many years.⁵ Often, the signs and symptoms are present but are misinterpreted as GERD. Patients frequently have heartburn, which is probably related to the food that eventually becomes acidic after retention in the esophagus. The acidity irritates the esophagus. A delay in diagnosing achalasia can be detrimental to the patient since early treatment may reduce the risk of developing cancer. Studies have shown that there may be a 16-

fold increase in cancer, specifically in squamous cell carcinoma.⁶ The role of endoscopy for cancer surveillance after achalasia treatment is unknown.

Once a diagnosis of achalasia is suspected, confirming the diagnosis is straightforward. The initial screening test is a barium swallow study. This will classically show a dilated distal esophagus with a smooth taper to the LES. The barium at this level will taper off and is often described as a “bird’s beak.” To confirm achalasia, esophageal manometry is the study of choice.² The findings will not only include an elevated resting LES pressure (typically greater than 45 mm Hg), but also incomplete relaxation of the esophagus during swallowing and absence of peristalsis.⁷ Endoscopy is performed prior to definitive treatment to rule out any disorder which may mimic achalasia, such as Chagas disease and cancer.

There are many options for the treatment of achalasia. The first therapy used by Sir Thomas Willis consisted of dilation using a sponge attached to a long, thin whale bone.⁸ Today, treatments for achalasia include medical management, endoscopic procedures, and surgery. Medical therapy consists of calcium channel blockers and nitrates.⁹ This option is unsatisfactory. Approximately 10% of patients will benefit from this, but often the side effects outweigh the benefits.^{10,11} Endoscopic procedures routinely used are botulin toxin injections and pneumatic dilation. The short-term results of these treatments have been shown to be excellent, but symptoms often return.¹² Endoscopic procedures may be appropriate in certain patients, but they can make more definitive treatment in the future difficult.¹³ Surgical treatment will provide the best long-term result,¹⁴ and newer techniques involving laparoscopic approaches have become the ideal treatments of choice because they shorten recovery time and length of hospital stay.¹⁵

The most common surgical treatment for achalasia is the Heller myotomy, first used by Ernest Heller in 1913. This technique involves cutting through the adventitia and both the longitudinal and circular smooth muscle layers of the muscularis propria. The cut is made down to the submucosa. The incision is extended from approximately 3–4 cm above the gastroesophageal junction to 2 cm onto the cardia of the stomach.¹⁶ There is some debate regarding whether to perform a partial fundoplication at this point. The most commonly used partial fundoplication is the Dor technique, in which the fundus of the stomach is brought anterior to the esophagus. Heller myotomy has been shown to decrease GERD, which is the most common complication following Heller myotomy, and to be more cost effective long-term in some studies.^{17–19} In this case, a fundoplication was not performed because of the very dilated esophagus.

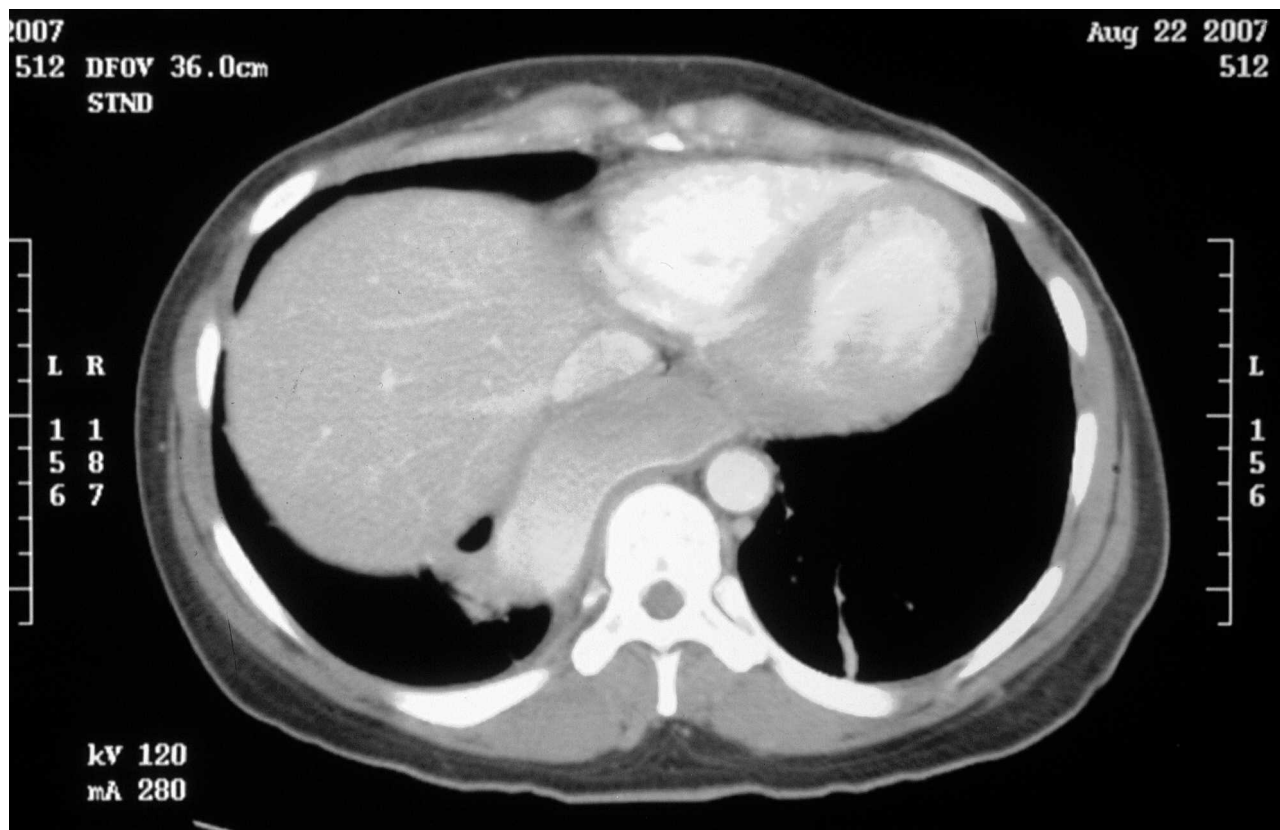


Figure 2. Representative image from the CT scan.

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