

Back Pain With A Congenital Cyst Of The Esophagus

Wm. David Huff, M.D., Gary Adams, M.D., and Walter J. Schoepfle, M.D.

Abstract: We report a 31-year-old man with a bronchogenic type of congenital esophageal cyst, which was discovered in the course of diagnostic evaluation for thoracic back pain, fever, and diarrhea. He was treated successfully by surgical excision of the cyst.

The embryology, classification, and symptoms of esophageal cysts are reviewed, along with useful diagnostic tests and procedures. Ordinary radiography, computed tomography, sonography, and magnetic resonance imaging contribute to diagnostic accuracy. Special mention is made of percutaneous fine needle aspiration biopsy (PNAB) as an adjunct diagnostic procedure.

Surgical excision is the treatment of choice. (J Am Bd Fam Pract 1989; 2:275-8.)

A 31-year-old man came to our office complaining of lower thoracic back pain for 3 days and fever (101 °F [38.3 °C]) for 1 day. Also, he said that he had diarrhea a week before the office visit, but it subsided after he had limited his diet to clear liquids for 24 hours.

He had had similar episodes of back pain in the last 2–3 years, but never for more than 1 day at a time. He acknowledged no other musculoskeletal or gastrointestinal complaints, but he reported a surgical history of tonsillectomy and inguinal hernia and hydrocele repair.

Physical Examination

He was well developed and nourished and did not appear to be distressed. His temperature was normal (98.6 °F [36 °C]), blood pressure (112/68 mmHg), and heart rate and rhythm were normal. There was no icterus, and his cardiopulmonary examination was normal. His abdomen was not tender, and no enlarged organs or masses were detected. His left testis was atrophied. There was no tenderness of his back, and the rectal examination showed no masses or occult fecal blood. There were no enlarged lymph nodes, and his deep tendon reflexes were present and symmetrical.

Laboratory Findings

Because of the history of diarrhea and recurrent back pain, flat and upright abdominal radio-

graphs to include a posterior-anterior chest view were obtained. The abdominal films were unremarkable, but the chest radiograph showed a right paraspinal mass extending from the level of T-9 to T-11, thought to be in the posterior mediastinum (Figures 1a and 1b). Computerized tomography (CT) scan confirmed the mass, which appeared to be solid (Figure 2). The patient was hospitalized for further evaluation. Initial CBC, urinalysis, serum electrolytes, and coagulation tests were normal. A CT-directed percutaneous fine needle aspiration biopsy of the mass failed to provide adequate tissue for diagnosis.

Treatment and Course

Thoracic surgical consultation was obtained, and surgical exploration through a right lateral thoractomy showed a cyst that was contiguous with the esophagus, displacing it to the left, but it did not communicate with the lumen of the esophagus. Pathological diagnosis was benign bronchogenic cyst. His postoperative course was uneventful. A postoperative water-soluble contrast study of the esophagus showed mildly decreased motility of the distal esophagus but was otherwise unremarkable.

On subsequent office visits, the patient's back pain was improved but still present, suggesting it was not definitely related to the cyst.

Discussion

Embryology

Congenital cysts of the esophagus are malformations that originate in the primitive foregut at the time of formation of the lower respiratory tract

From the School of Primary Medical Care, University of Alabama, Huntsville. Address reprint requests to Wm. David Huff, M.D., 600 St. Clair, #9, Huntsville, AL 35801.

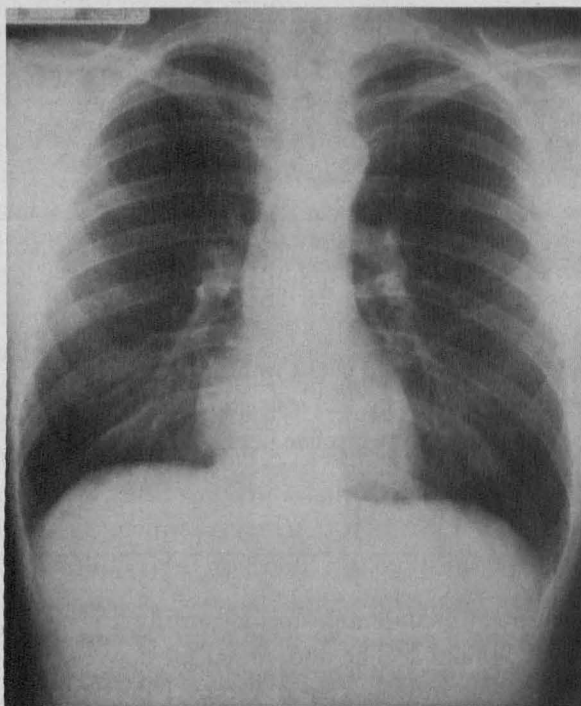


Figure 1a. PA chest radiograph with right paraspinal mass (shown faintly).

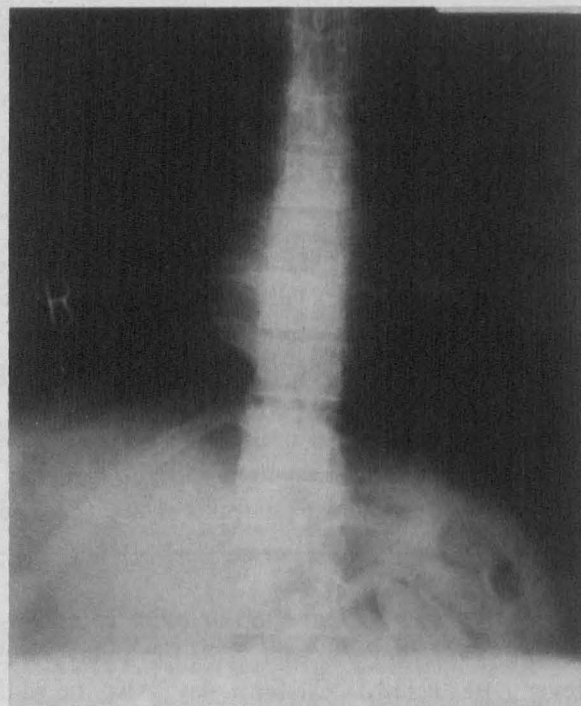


Figure 1b. Coned-down view of chest radiograph showing right paraspinal mass.

and the stomach.¹ After leiomyoma, they represent the second most common benign esophageal tumor² and may be lined with gastric, enteric, or respiratory epithelium.³ There are several embryological theories about the development of esophageal cysts. The most popular theory was proposed by Bremmer in 1944.⁴ The foregut of the embryo is covered by cells similar to respiratory epithelium. These cells produce secretions that form vacuoles; subsequently, they orient longitudinally and coalesce to form a lumen.¹⁴ If for any reason some vacuoles fail to coalesce in the longitudinal axis, a cyst will form and then become surrounded by muscle layers.

There seems to be a 2:1 male predominance of these duplication cysts, with approximately 50 to 60 percent located in the lower third of the esophagus. A review of 49,196 autopsies reported only 6 esophageal cysts, a frequency of 1 per 8200.⁵

Congenital esophageal cysts are classified as *duplication* cysts, *bronchogenic* cysts, *gastric* cysts, and *inclusion* cysts.⁵ Duplications are within the esophageal wall, covered by two muscle layers, and contain squamous epithelium or a lining

compatible with that found in the embryonic esophagus. A bronchogenic cyst is located intramurally and contains cartilage. A gastric cyst must be found within the esophageal wall, contain one or more muscle walls, and be lined with gastric mucosa. Inclusion cysts are intramurally located and contain epithelium that is of a respiratory or squamous type. They are not covered by two muscle layers, nor do they contain cartilage; therefore, they cannot be called duplication or bronchogenic cysts.

Acquired cysts of the esophagus may form from glands in the mucous and submucous layers of the esophagus. The reason for this formation is not known.

Clinical Symptoms

The symptoms caused by esophageal cysts may be varied. In adults, the lesions are frequently asymptomatic.⁶ If the cyst has become large, dysphagia, regurgitation, vomiting, anorexia, weight loss, and pain may develop because of pressure on the esophagus. Symptoms of tracheobronchial pressure include coughing, dyspnea, wheezing, choking episodes, and recurrent respiratory infections.

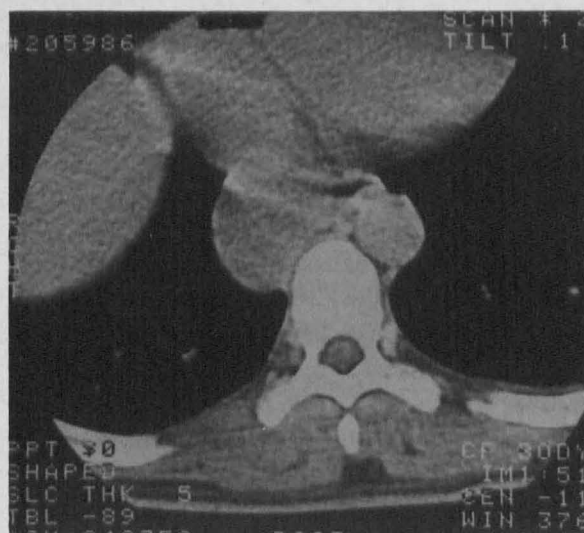


Figure 2. CT scan of chest showing right paraspinal mass.

Gastric cysts may have symptoms of peptic ulcer disease and may, in fact, bleed or hemorrhage. Occasionally, esophageal duplications are associated with neurologic deficits, limitation of motion of the neck, scoliosis, root compression, or pain.⁷

Diagnosis and Treatment

The differential diagnosis of an esophageal cyst includes all intramural extramucosal tumors of the esophagus: leiomyoma, lipoma, fibrolipoma, neurinoma, and neurofibroma. The chest radiograph remains the primary initial diagnostic examination.⁸ This may provide information about anatomical location, size, and degree of calcification. Barium swallow and sonography may also aid in the diagnosis.

CT scan and magnetic resonance imaging (MRI) are also useful in the evaluation of mediastinal masses to determine size, location, solid or cystic structure, and relation to other structures. However, it cannot be determined that a mass is malignant or benign on the basis of these studies. Malignant degeneration of esophageal cysts has been described⁹; therefore, surgical excision is the treatment of choice for congenital esophageal cysts.

The short-term results after surgical removal of congenital esophageal cysts are excellent. However, a recent study of long-term follow-up showed a 64 percent occurrence of reflux symp-

toms and esophagitis as documented by endoscopy.¹⁰ The etiology of the esophagitis is unclear, but this finding does indicate the need for long-term monitoring of all patients with esophageal cysts who are treated surgically.

Another useful diagnostic tool in evaluation of mediastinal masses is percutaneous fine needle aspiration biopsy (PNAB).¹¹ The cytological diagnosis of mediastinal metastases from lung and extrathoracic sites and the evaluation of cysts are very accurate via the PNAB technique. It is important to attempt PNAB before surgery, because it is especially helpful in the cytological diagnosis of thymoma. A thymic lesion is the second most frequent mediastinal mass¹² and the most common anterior mediastinal tumor.¹³ It is important to make the diagnosis of thymoma preoperatively because if the tumor is very large or is suspected of being malignant, a preoperative course of radiotherapy and chemotherapy is advisable.⁹

The risk from PNAB is minimized if a vascular etiology of an intrathoracic mass is first excluded. The use of CT scanning, sonography, or MRI aids in this distinction.

The authors thank Dr. William J. Crump, Associate Professor of Family Medicine, School of Primary Medical Care, Huntsville, Alabama, for his review and suggestions. Also, a special thanks to Ms. Clarice Thomas for her efforts and patience in the preparation of this manuscript.

References

1. Latimer RD. Enterogenous cysts of the oesophagus. *Br J Dis Chest* 1967; 61:136-7.
2. Anderson HA, Pluth JR. Benign tumors, cysts, and duplications of the esophagus. In: Payne WS, Olsen AM, eds. *The esophagus*. Philadelphia: Lea & Febiger, 1974:225-37.
3. Boyd DP, Hill LD. Benign tumors and cysts of the esophagus. *Am J Surg* 1957; 93:252.
4. Bremer JL. Diverticula and duplications of the intestinal tract. *Arch Pathol Lab Med* 1942; 38:132-40.
5. Arbona JL, Fazzi JG, Mayoral J. Congenital esophageal cysts: case report and review of literature. *Am J Gastroenterol* 1984; 79:177-82.
6. Postlethwait RW. *Surgery of the esophagus*. Norwalk, CT: Appleton-Century-Crofts, 1979.
7. Sethi GK, Marsden J, Johnson D. Duplication cysts of the esophagus. *South Med J* 1974; 67:616-8.
8. Davis RD, Oldham HN, Sabiston DC Jr. Primary cysts and neoplasms of the mediastinum: recent

- changes in clinical presentation, methods of diagnosis, management and results. *Ann Thorac Surg* 1987; 44:229-37.
9. Bergman M, Charnos RM. Tracheobronchial rests in the esophagus: their relation to benign structures and certain types of cancer of the esophagus. *J Thoracic Surg* 1968; 35:97.
 10. Salo JA, Ala-Kulju KV. Congenital esophageal cysts in adults. *Ann Thorac Surg* 1987; 44:135-8.
 11. Weisbrod GL, Lyons DJ, Tao LC, Chamberlain DW. Percutaneous fine-needle aspiration biopsy of mediastinal lesions. *AJR* 1984; 143:525-9.
 12. Robison RI, Pavlina PM, Scherer LR, Grosfeld JL. Multiple esophageal duplication cysts. *J Thorac Cardiovasc Surg* 1987; 94:144-7.
 13. Fontenelle LJ, Armstrong RG, Stanford W, Lindberg EF, Dooley BN. The asymptomatic mediastinal mass. *Arch Surg* 1971; 102:98-102.

Editorial Comment

The two preceding case reports, which involve interesting chest lesions, were submitted for

publication independently. They both demonstrate the need for careful presurgical evaluation.

The case of the congenital cyst of the esophagus discusses the potential value of percutaneous fine needle aspiration biopsy in suspected solid or cystic lesions. However, the other case represents the problem of a mediastinal vascular lesion. In this case, angiography is recommended and presumably PNAB may be quite hazardous.

Both cases show the need for careful radiographic study prior to invasive procedures. Precise and accurate interpretations of noninvasive diagnostic information is of critical importance to assure a positive outcome.

Although each case is independently instructive, taken together, the educational value seems enhanced.

Paul R. Young, M.D.
Lexington, KY

American Board of Family Practice Certification/Recertification Examination Dates

July 13, 1990
July 12, 1991
July 10, 1992
July 9, 1993
July 8, 1994