THE PATIENT
36-year-old man

SIGNS & SYMPTOMS
– Persistent dry cough
– Frequent sinus congestion
– Hemoptysis

Rade N. Pejic, MD, MMM; Andrew T. Gabrielson, MD
Department of Family Medicine, Tulane University, New Orleans, LA (Dr. Pejic); James Buchanan Brady Urological Institute, Johns Hopkins University School of Medicine, Baltimore, MD (Dr. Gabrielson)

rpejic@tulane.edu

The authors reported no potential conflict of interest relevant to this article.

THE CASE

A 36-year-old nonsmoking white man presented with an episodic 3-month history of dry cough and nasal allergy symptoms. He reported a past history of sinus allergies but no history of asthma. His illness began with a flu-like syndrome, and he had been treated with antibiotics (amoxicillin and azithromycin) and oral steroids (methylprednisolone) by 2 other physicians for "viral syndrome" and "bronchitis."

The patient reported some tactile fever initially but none thereafter. Symptoms included episodic wheezing but no overt shortness of breath. In addition to the persistent dry cough, he complained of frequent sinus congestion, post-nasal drip, and sneezing. He became concerned when he noticed a fleck of blood in his phlegm.

Physical exam was unimpressive, except for nasal congestion. His breath sounds were clear. Chest x-ray showed a benign-appearing granuloma in the right lower lobe (no previous films available for comparison). Peak-flow measurements taken in the office were persistently low (58%-70%) but improved with steroids and inhaled albuterol.

Over the following 7 weeks, the patient experienced waxing and waning symptoms. At his follow-up visit, he appeared well; chest auscultation revealed normal breath sounds. He was treated with an additional round of antibiotics (levofloxacin), oral steroids, nasal steroids, and inhaled albuterol.

At 13 weeks from his initial presentation, he developed frank hemoptysis and was diagnosed with a right lower-lobe pneumonia in the emergency department. While hospitalized, his clinical status deteriorated, requiring chest tube placement for a large pleural effusion.

Shortly thereafter, he underwent right middle and lower lobectomies and decortication. Multiple organisms were cultured from the pleural fluid. Tuberculosis testing and acid-fast bacilli stains were negative. No malignant cells were identified. Pathologic examination of the resected lung tissue confirmed the chest x-ray finding of a benign calcified granuloma. Additional testing, including a thin barium esophagram, was performed.

THE DIAGNOSIS

Results of the esophagram revealed a congenital bronchoesophageal fistula (C-BEF) between the patient's esophagus and right mainstem bronchus, located 15 cm distal to his trachea.

DISCUSSION

Fistulous connections between the esophagus and bronchi are rare but may arise in the setting of malignancy, trauma, inflammation, or congenital malformation.[1] While the precise etiology of C-BEF remains unknown, it is believed to be a consequence of failed tracheoesophageal separation during the early stages of embryonic development.

Prevalence and epidemiology. C-BEF has been reported to occur in 1 in 3000 to 4000 live births, often with concomitant esophageal atresia.[2] Infants with esophageal atresia

demonstrate clinically significant respiratory symptoms and failure to thrive. However, C-BEF without esophageal atresia may be asymptomatic for years to decades.

Age at diagnosis ranges from 9 days to 83 years.[3] Several explanations exist for the prolonged asymptomatic phase of this disease: (1) presence of a membrane overlying the fistula during childhood that subsequently ruptures; (2) presence of a proximal fold of esophageal mucosa overlapping the orifice; (3) antigravitational or upward extension of the fistulous tract from the esophagus; and (4) spasm of the smooth muscle of the fistula.[4]

Four subtypes. Type I fistulas are associated with a wide-necked congenital diverticulum of the esophagus, which may become inflamed and allow perforation into the nearby lung. Type II fistulas (most common) consist of a short tract running directly from the esophagus to a nearby lobar or segmental bronchus. Type III fistulas involve a communication between the esophagus and a cystic structure within the lung parenchyma. Type IV fistulas run from the esophagus into a sequestered pulmonary segment.[1] Our patient had a type II fistula.

Congenital bronchoesophageal fistula without esophageal atresia may be asymptomatic for years to decades.

Is there a nonspecific cough? The most common signs and symptoms of C-BEF are nonspecific cough, cough after ingestion of fluids or meals, and hemoptysis.[5,6] Symptoms may persist for decades prior to diagnosis, and the indolent course of C-BEF may lead to fatal complications such as recalcitrant pneumonia, bronchiectasis, and abscess formation.

One test bests others for diagnosis. Plain chest x-ray may indicate enlarged lymph nodes or surrounding airspace disease but will not be able to identify a C-BEF. Computed tomography (CT) of the chest may detect the presence of a C-BEF but does not rule it out. Barium esophagram is the most sensitive test for BEF. Esophagoscopy and bronchoscopy may be helpful once the BEF has been identified, but neither has demonstrated reliability as a first-line test. In this particular case, the C-BEF was not seen on chest CT but was later detected on a thin barium esophagram.

To confirm the congenital nature of the fistula, histopathology should be examined. C-BEFs will have a mucosal layer and definitive muscularis layer within the fistulous tract.[3,5]

Treatment. The preferred method of treatment for C-BEF is thoracotomy with resection of the fistula and insertion of a pleural or muscular flap graft to close the defects in the bronchus and esophagus.[7] Alternatively, obliteration of the esophageal defect can be performed using biological glue or silver nitrate. Prognosis after surgical repair is excellent.

Our patient

Two weeks after hospital discharge, the patient was re-admitted for hydropneumothorax and underwent additional surgeries. Unfortunately, he died in the ICU due to a tension pneumothorax while intubated.

THE TAKEAWAY

C-BEF is a rare, insidious condition that may remain asymptomatic into adulthood. After common causes are ruled out, patients with adult-onset nonspecific cough, episodes of coughing after eating/drinking, and hemoptysis should be evaluated for BEF. The most useful diagnostic investigation is barium esophagram. Once C-BEF is identified, prompt surgical management is warranted. Because C-BEF persisting into adulthood is so rare, recommendations regarding diagnosis and treatment are based on expert opinion. JFP

CORRESPONDENCE
Rade N. Pejic, MD, MMM, Department of Family Medicine, Tulane University School of Medicine, 1430 Tulane Avenue, Mailbox #8033, New Orleans, LA 70112; rpejic@tulane.edu

REFERENCES

1. Braimbridge MV, Keith HI. Oesophago-bronchial fistula in the adult. *Thorax*. 1965;20:226-233.
2. Taira N, Kawasaki H, Atsumi E, et al. A rare case of congenital bronchoesophageal fistula in an adult. *Int J Surg Case Rep*. 2017;36:182-184.
3. Risher WH, Arensman RM, Ochsner JL. Congenital bronchoesophageal fistula. *Ann Thorac Surg*. 1990;49:500-505.
4. Paul M, John S, Ashton R. Recurrent pneumonia in a 51-year-old woman due to congenital bronchoesophageal fistula. *Respir Care*. 2011;56:1203-1205.
5. Rämö OJ, Salo JA, Mattila SP. Congenital bronchoesophageal fistula in the adult. *Ann Thorac Surg*. 1995;59:887-889.
6. Zhang B-S, Zhou N-K, Yu C-H. Congenital bronchoesophageal fistula in adults. *World J Gastroenterol*. 2011;17:1358-1361.
7. Su L, Wei X-Q, Zhi X-Y, et al. Congenital bronchoesophageal fistula in an adult: a case report. *World J Gastroenterol*. 2007;13:3776–3777.