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Eventration of the diaphragm presenting as spontaneous pneumothorax

A 25-YEAR-OLD MAN with a 2-day history of upper respiratory tract infection presents to the emergency department with the sudden onset of right-sided back and chest pain and shortness of breath after a severe coughing fit.

He is morbidly obese, is a long-time smoker, and has had recurrent exacerbations of asthma with frequent upper respiratory tract infections. He has no history of recent trauma.

A review of systems reveals no significant impairment in exercise tolerance. He has been able to continue doing manual labor at his job as a railroad worker.

Radiography shows a large right pneumothorax and an elevated right diaphragm (**Figure 1**). Computed tomography (CT) (**Figure 2**) reveals a right anterior apical pneumothorax with hypoplastic lung and significant elevation of the right diaphragm with fat, bowel, and kidney within the right thorax. He is hemodynamically stable and shows no signs of bowel obstruction.

The physical examination is normal except for diminished breath sounds on the right side. He is diagnosed with congenital diaphragmatic hernia and spontaneous pneumothorax. A 10-F locking pigtail catheter is inserted under CT guidance, leading to complete resolution of the pneumothorax. He is discharged home the next day with a plan for elective repair of the hernia.

Two months later, he returns for scheduled right thoracotomy to repair the hernia. However, while preparing the chest cavity, the surgeon finds no diaphragmatic hernia and no intra-abdominal content—but rather, a severely elevated and thinned-out diaphragm with uninterrupted continuity. The diagnosis is changed to congenital diaphragmatic eventration, and plication of the diaphragm is performed with a series of interrupted, pledgeted polypropylene sutures.



FIGURE 1. Plain radiography shows a large right pneumothorax and an elevated right diaphragm.

CONGENITAL EVENTRATION OF THE DIAPHRAGM

Congenital diaphragmatic eventration is a rare developmental defect of the central, muscular portion of the diaphragm. The true prevalence is not known, but early reports identified this condition in less than 0.1% of adults.¹

Symptomatic patients usually experience dyspnea secondary to ventilation-perfusion mismatch resulting from chronic atelectasis and lung hypoplasia, as well as impaired ventilation resulting from the limited caudal migration of the diaphragm.^{2,3} Increased susceptibility to recurrent upper respiratory tract infections and pneumonia is also a common feature.

Although rare, spontaneous pneumothorax can develop in patients such as ours, whose lengthy history of smoking and asthma predisposed him to the development of emphysema-like blebs and bullae and to subsequent rupture of blebs brought on by vigorous

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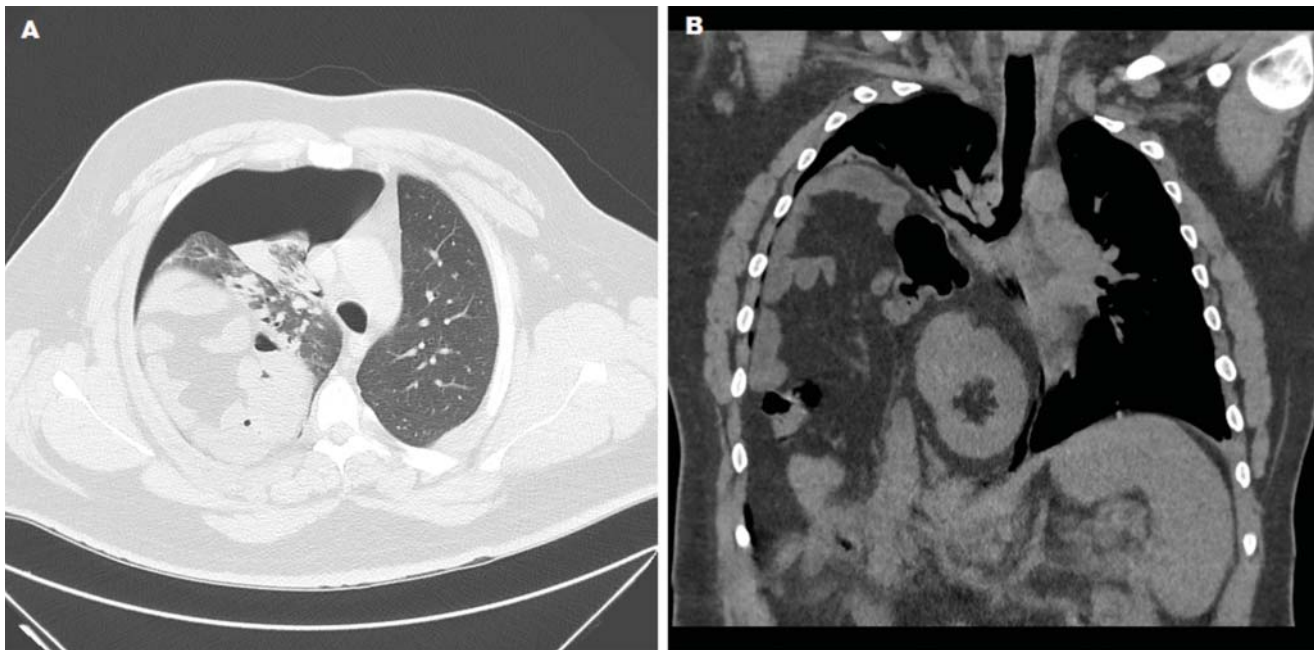


FIGURE 2. Thoracic computed tomography shows right anterior apical pneumothorax with atelectatic and hypoplastic lung in a cross-sectional view (A). A coronal view (B) shows significant elevation of the right hemidiaphragm, with fat, bowel, and kidney within the right hemithorax.

Before considering surgery, rule out other causes of symptoms

coughing that caused an involuntary Valsalva maneuver.⁴

As in our patient, distinguishing congenital diaphragmatic eventration from hernia preoperatively can be difficult with plain chest radiography. Spiral CT with multiplanar reconstruction or with magnetic resonance imaging can help establish the diagnosis.³ However, a severely attenuated diaphragm can be difficult

to visualize on CT, as in our patient, leading to a presumptive diagnosis of diaphragmatic hernia. In such situations, the diagnosis of eventration can only be made intraoperatively.

Surgical repair is indicated only for patients with symptoms. Other potential causes of the symptoms should first be ruled out, however, including primary pulmonary disease, cardiac dysfunction, and morbid obesity. ■

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