Clinical-Pathologic Conference

Clinical-pathologic conference in general thoracic surgery: Malignant transformation of recurrent respiratory papillomatosis

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Case Presentation

Dr Kozower: A 30-year-old woman presented with recurrent left-sided pneumonia and a left lower lobe lung abscess. She worked as a nurse but had been on disability for 6 months because of continuous purulent sputum production, fever, weight loss, and fatigue. Her past medical history is significant for recurrent respiratory papillomatosis, which has required numerous procedures to maintain a patent airway, including a tracheostomy performed in 1977. A previous left thoracotomy was performed at an outside hospital, but the procedure was abandoned because of technical difficulties with ventilation. She was referred here for evaluation and possible left lower lobe lobectomy.

She has no smoking history and does not consume any alcohol. She has 5 siblings, all of whom are in good health. She has no other significant past medical history. Her surgical history includes the tracheostomy performed at age 3 years and multiple bronchoscopies and airway interventions to maintain patency. On physical examination, she was very thin but appeared well. Her stoma was clean, with no signs of infection. She had no cervical or supraclavicular adenopathy, and her pulmonary examination was only significant for decreased breath sounds over the left base. The left thoracotomy incision was well healed. Bronchoscopic biopsy specimens from a large papilloma in her left lower lobe were performed at an outside hospital. The earliest was in April 2004 and showed some benign tracheal mucosa and papilloma. She had another biopsy 2 months later that showed some atypical cells in the left lower lobe bronchus.

Dr Javidan: The chest radiograph shows a tracheostomy tube, mediastinal widening suggestive of mediastinal lymphadenopathy, a left pleural effusion, and multiple nodules and masses in both lungs (Figure 1). Most of the masses are cavitated, and many have air fluid levels within them. These lesions are better evaluated with computed tomography (CT). CT shows multiple cavitating nodules and cystic lesions with varying degrees of wall thickening (Figure 2). These lesions are characteristic for adults with papillomatosis of the lungs.

In this patient the soft-tissue windows of the CT scan show a mass invading the left lower lobe bronchus, accompanied by distal segmental collapse of the postero-

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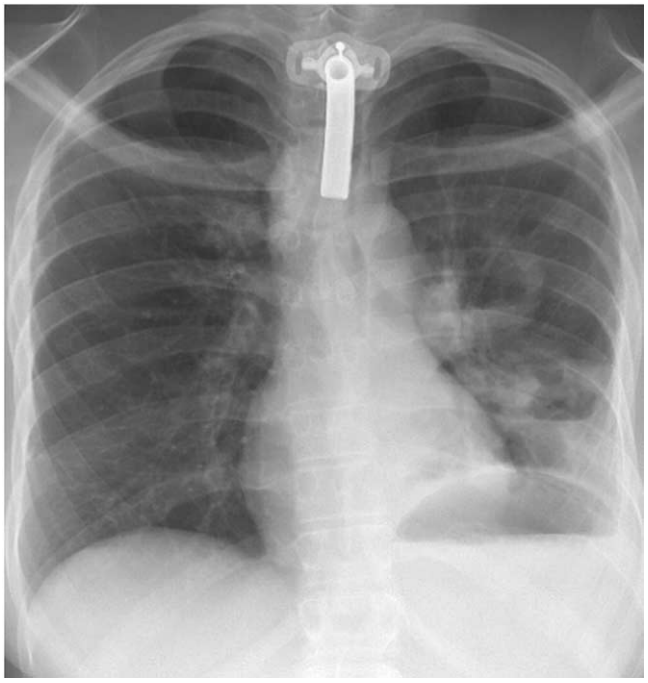

Figure 1. Posteroanterior upright chest radiograph showing multiple cavitary masses containing fluid in the left lower lobe, a right suprahilar mass, a left pleural effusion, mediastinal widening, and left hilar enlargement, which is suggestive of lymphadenopathy.

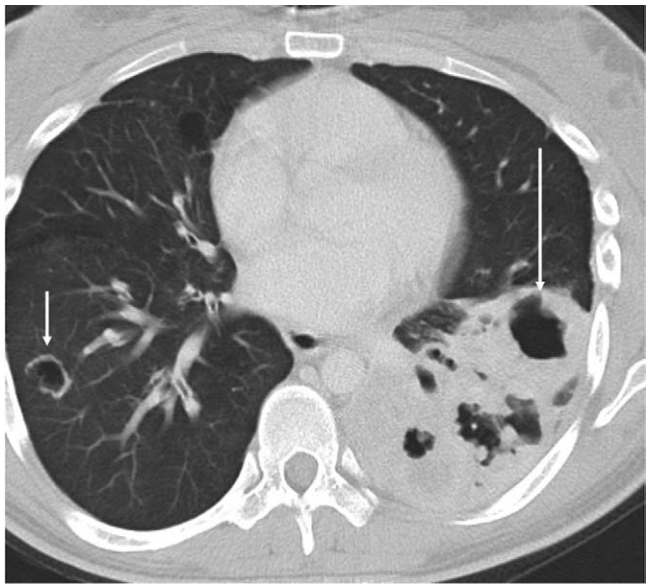

Figure 2. Computed tomographic lung window image shows a thinner-walled cystic lesion in the right lower lobe *(short arrow)* and a larger fluid-containing cavitary lesion in the left lower lobe with an irregular thick wall. Segmental collapse of the posterobasal segment of the left lower lobe is accompanied by more cavitary lesions.

basal segment of the left lower lobe and multiple cavitary lesions with irregular thick walls and mural nodules in the remainder of the left lower lobe (Figure 3). The soft-tissue mass and the cavitary lesions could represent papillomatosis or malignant transformation into squamous cell carcinoma.

In the left lower lobe there are some air- and fluid-filled dilated bronchi and bronchioles, which are due to postobstructive atelectasis in the setting of the slow-growing central mass growing into the left lower lobe bronchus. There are multiple enlarged lymph nodes in the mediastinum, many of which have centers of lower attenuation. This lymphadenopathy could represent superimposed infection, an abscess, human papilloma virus (HPV) infection, or metastatic lymphadenopathy resulting from malignant transformation.

There is a broad differential diagnosis for multiple, cavitating, thick-walled lesions. First is squamous cell cancer, primary or metastatic. HPV, Wegener's granulomatosis, and fungal infections are other causes. Because of this patient's history of respiratory papillomatosis and the risk for malignant transformation in the airways and lung parenchyma, I am most worried about squamous cell cancer. The lymphadenopathy is concerning but might be reactive to the HPV infection or lung abscess.

Dr Patterson: Bear in mind that this patient had a tracheostomy performed when she was 3 years of age because of tracheal papillomatosis, and she has had hundreds, maybe thousands, of bronchoscopic debridements, laser, cautery, fulguration, etc. Her symptoms were chronic illness, continuous production of purulent sputum, constant coughing, and weight loss. All of her symptoms were attributed to that cavity in the lower lobe. In addition, she had a large papillomatous obstruction of her left lower lobe. About 6 months before coming here, she had a thoracotomy with the intent of removing that left lower lobe abscess or cavity. During the conduct of that operation, the patient had an episode of desaturation, and the surgeons abandoned the procedure, closing the thoracotomy. She was subsequently transferred here.

Dr Cooper: Was her larynx obstructed?

Dr Patterson: Yes, her upper airway was obliterated by a combination of scar and papillomatosis.

Dr Kozower: On December 6, we performed flexible and rigid bronchoscopy, which revealed extensive purulent secretions coming from both lungs. After these were suctioned, we determined the focus to be in her left lower lobe. In addition, there was a large papilloma seen in the left lower lobe, with a good cuff of proximal airway, and we thought that she would be a candidate for left lower lobe lobectomy. Given the enlarged lymph nodes that were seen