A Patient With Shortness of Breath and a Left Hilar Mass

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A 38-year-old African American man with a history of smoking was referred to our medical center by his family physician for shortness of breath and abnormalities on chest x-ray film. He was compelled to resign his position as a concrete worker because of increasing shortness of breath during the previous 8 weeks. Tuberculosis was suspected but ruled out after results of sputum acid-fast stains and cultures were negative. Thoracic computed tomography scan revealed a mass in the left chest encircling the left mainstem bronchus. Bronchoscopic examination showed a large mass nearly obstructing the left mainstem bronchus. Bronchial brush and bronchial wash samples were submitted for cytologic assessment and showed predominantly benign bronchial cells and macrophages as well as a few, small, blue, atypical cells of uncertain histogenesis. Submitted biopsies were nondiagnostic showing predominantly necrotic tissue with fibrin and fragments of bronchial mucosa with papillary and squamous metaplasia. Because of clinical symptomatology and the likelihood of malignancy, a left lower lobectomy was performed. In addition, lymph nodes from various peripulmonary sites were submitted. The left lower lung lobe was submitted fresh, and frozen sections of the bronchial resection margin and a subcarinal lymph node were negative for malignancy. Gross examination showed a 590-g left lower lobe of lung with multiple attached hilar lymph nodes. The visceral pleura had a tan-gray and light brown mottled appearance. Sectioning of the lung revealed a spheroid, well-circumscribed, fleshy tumor in the superior portion of the lung lobe (Figure 1). The tumor measured $8 \times 5 \times 4.5$ cm and the cut surface was soft and tan-brown with focal areas of cystic degeneration. Approximately 30% of the tumor was grossly necrotic. The tumor was within 5 mm of the anterior pleural surface and grossly invaded the parenchyma beneath the main bronchus near the surgical resection margin. The remainder of the lung showed extensive consolidation and purulent mucus plugs in the bronchial tree. Hematoxylin-eosin-stained sections of the tumor showed neoplastic glands arranged in a stroma composed of a focally pleomorphic population of small, oval- and spindle-shaped cells lying in a myxoid matrix with crowding around the neoplastic glands. Neoplastic glands morphologically resembled endometrial glands (Figures 2 and 3). Frequent mitoses and prominent areas of hemorrhage and necrosis were seen. Bronchial and pulmonary artery margins were negative for tumor involvement. Submitted lymph nodes showed sinus histiocytosis but no evidence of metastatic tumor.

What is your diagnosis?

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Pathologic Diagnosis: Biphasic Pulmonary Blastoma

Abstract

Our patient, a 38-year-old African American man with a history of smoking, presented with shortness of breath and a left hilar mass on imaging studies. A left lower lobectomy was performed that demonstrated a grossly well-demarcated, fleshy, tan-brown tumor that measured 8 cm in greatest dimension. Microscopic examination revealed neoplastic glands with an endometrioid morphology surrounded by a myxoid stroma containing focally pleomorphic, small, oval- and spindle-shaped cells. Frequent mitoses and prominent areas of hemorrhage and necrosis were noted. Margins of resection and lymph nodes were negative for tumor. A diagnosis of biphasic pulmonary blastoma was rendered. The patient had an uneventful postoperative course and was discharged in stable condition 6 days following the procedure.

Pulmonary blastoma is a rare tumor of lung composed of neoplastic mesenchymal and epithelial components that histologically resemble embryonal lung; a biphasic morphology was seen in this case. The tumors have a poor prognosis and a variable clinical course that cannot be determined from histologic appearance.

According to the 2004 World Health Organization histologic classification of lung tumors, pulmonary blastoma, also referred to historically and in the literature as biphasic pulmonary blastoma (BPB), falls under the heading of sarcomatoid carcinomas. Although BPB contains both a primitive-appearing epithelial component that often resembles well-differentiated fetal adenocarcinoma (WDFA) and also a mesenchymal component that may resemble pleuropulmonary blastoma, these other entities have been classified under the categories of adenocarcinomas and mesenchymal tumors, respectively, and are distinctive clinical, morphologic, and molecular entities. BPBs consist of relatively equal proportions of malignant epithelial and mesenchymal components. A predominance of malignant epithelial elements characterizes WDFA, which has a better prognosis than BPB. Pleuropulmonary blastoma is a rare primary embryonal mesenchymal malignant neoplasm arising in pulmonary parenchyma, in association with pleura, or within the mediastinum. BPB and WDFA occur only in the lung parenchyma and generally affect adults in the fourth or fifth decade with occasional presentation in children. Pleuropulmonary blastoma is found in children, usually younger than 3 years.

Grossly, the tumors are large, well-demarcated but unencapsulated solitary peripheral masses. The sizable tumor in this case compressed bronchial branches, resulting in distal bronchiectasis, as seen in Figure 1. Histologically, BPB has a primitive epithelial component that may resemble WDFA and is composed of tubules, acini, or a combination and contains an epithelial component corresponding to low grade adenocarcinoma of the fetal lung type/WDFA or classic pulmonary blastoma with complex glandular structures resembling fetal lung and morules with biotin-rich optically clear nuclei. The second has a high-grade adenocarcinoma of fetal lung type/clear cell adenocarcinoma with fetal lung features as its epithelial component and occurs predominantly in elderly men who are heavy smokers. This tumor has been termed blastoma-like or blastomatoid variant of carcinosarcoma. These entities may be distinguished by immunohistochemistry to detect aberrant nuclear/cytoplasmic localization of β-catenin protein due to β-catenin gene mutations. β-Catenin overexpression in both epithelial and mesenchymal components, especially in budding glands and morules, may be characteristic of the WDFA-related variant.

Shortness of Breath and Hilar Mass—Kaibipour & Lankachandra