Adenoid Cystic Carcinoma of the Lung with Bilateral Renal Metastases at Presentation in a Young Female

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Abstract

Adenoid cystic carcinoma of the lung is a relatively rare, slow growing lung neoplasm. Metastases outside the lung are uncommon. Herein, we have reported the case of a patient who presented with a large mass in the right lower lobe of her lung. Bronchial biopsy revealed features suggestive of adenoid cystic carcinoma of the lung with a predominant cribriform architecture. CT abdomen showed features of bilateral renal and liver metastases, but no adrenal metastases.

Keywords: Adenoid cystic carcinoma of the lung, Renal metastasis, Cribriform pattern.

Introduction

Unusual tracheobronchial tumors represent 4% to 6% of all lung tumors (benign and malignant). Adenoid cystic carcinoma (ACC) accounts for less than 0.2% of all lung tumors. Pulmonary ACC usually arise in the proximal tracheobronchial tree, and are regarded as slowly growing tumors. Metastases outside the lungs, such as the kidneys, are infrequent and occur much later after initial presentation. Herein, we have reported a case of primary ACC of the lung in a young female who presented with metastases to the liver and both kidneys.

Case Report

A 28-year-old female, non-smoker, was admitted with complaints of persistent progressive dyspnea, abdominal pain for one month along with two episodes of hemoptysis. No acid fast bacilli were found on sputum examination. Chest X-ray showed a large homogenous opacity in the right lower lung field. CT contrast scan of the thorax showed a heterogenous large mass (8.1 6.1cm) in the right lower lobe that was merged with an enlarged right hilar lymph node (Figure 1). Right paratracheal, pretracheal, and right hilar lymphadenopathy was seen with inhomogenous enhancement. CT-guided fine needle aspiration biopsy (FNAB) of the mass showed mucoid globules surrounded by medium-sized
epithelial cells, monomorphous in appearance, suggestive of ACC. On fiberoptic bronchoscopy, a proliferative growth was found on the right intermediate bronchus situated >2 cm from the carina. Bronchial brush cytology smear showed round cells in clusters; microacini showed hyperchromatic nuclei suggestive of a neoplastic epithelial lesion. Bronchial biopsy confirmed a tumor that was composed of epithelial cells arranged in a cribriform architecture, nests, and trabeculae. These cells were basaloid with predominantly monomorphic round hyperchromatic nuclei. Glandular spaces contained either eosinophilic or basophilic mucinous material. A diagnosis of ACC of the lung was made (Figure 2). On immunohistochemistry, tumor cells were positive for keratin and TTF-1, which established the diagnosis of ACC. No abnormality was detected in the salivary glands and thyroid on detailed investigations. CT abdomen noted multiple metastatic lesions in the liver with multiple metastatic space occupying lesions in both kidneys, but no adrenal metastases (Figure 3). In conclusion, a diagnosis of primary pulmonary ACC of the lung with metastases to both kidneys and the liver was made. The patient underwent a chemotherapy regimen that consisted of cisplatin and doxorubicin. However, after three cycles of chemotherapy, the patient died.

Discussion

Adenoid cystic carcinoma, also called cylindroma, is a distinctive malignant tumor that can arise from submucosal glands of the respiratory tract. The most common involved site in the respiratory tract is the trachea; bronchial lesions are very uncommon. Primary pulmonary ACC is an unusual cause of lung cancer, accounting for less than 0.2% of cases. The mean age of occurrence is 44.8 years with a male:female ratio of 1:1.1. A variety of presentations have been noted, such as: breathlessness (72%), wheezing (39%), cough (23%), stridor (21%) and hemoptysis (18%). Our case was atypical because the patient was only 28 years old.

Primary pulmonary ACC has a unique histology of submucosal and perineural infiltration and often extends 1 cm or more beyond the
Adenoid Cystic Carcinoma of the Lung with Bilateral Renal macroscopic margins. There are three histological subtypes: cribriform (most frequent), tubular, and solid, which is associated with an aggressive clinical course and distant metastases. The cribriform pattern is the most characteristic and recognizable form. It manifests as a punched-out or 'Swiss-cheese' arrangement of tumor cells, usually surrounding acellular spaces that may contain mucoid or hyaline material. Characteristic histopathological features of ACC allow for diagnosis based on hematoxylin-eosin stained biopsy specimens. Immunohistochemistry is also required to distinguish ACC from adenocarcinoma as ACC stains positively with keratin, actin, and vimentin. Adenoid cystic carcinoma of the lung usually occurs due to metastases from other sites. Thus, immunostaining for TTF-1 is required to differentiate primary from secondary lung cancers. In our patient, immunohistochemistry was positive for TTF-1 and keratin. Therefore, the diagnosis of primary pulmonary ACC was established.

This is a slow growing, low-grade malignancy that metastasizes to the lungs. Metastases outside the lungs are rare and occur late (in one series 12–300 months, mean 100 months), even three decades after initial presentation. The most common sites for distal metastases are the liver, bones, and brain. Renal metastases are unusual from ACC of the lungs.

The reported case has shown that pulmonary ACC may have a very aggressive behavior. Although ACC occurs in the salivary glands, ACC of the lung is rare. The key to cytology is a globule of mucous. ACC should be suspected when atypical small cells conglomerate even when the globule of mucus is absent. A correlation is known to exist between clinical presentation of ACC and histological subtypes. The solid histological pattern has been associated with a more aggressive clinical course and early distant metastases, in contrast to the cribriform type, which shows a more benign behavior. In our case, however, the cribriform subtype-predominant pattern resulted in very aggressive clinical course. Clinically recognized metastases to the kidney from lung carcinomas are rare despite the fact that renal metastases from lung primaries are

Figure 2. Histopathological section of bronchial biopsy showing tumor composed of epithelial cells arranged in a cribriform architecture, nests and trabeculae (H&E 200×)
frequent at post mortem examination. The kidney is a rare site of metastatic disease from primary tumors of the lung and is the fifth most common site of metastases in the body after the lungs, liver, bones, and adrenals. As many as 40% of renal metastases due to lung cancer are bilateral. Metastatic renal diseases are seldom clinically identified because the symptoms of pain and hematuria occur in only 20% of patients. Isolated renal metastases without adrenal metastases in lung cancer is very rare. Only 10% of renal metastases in lung cancer patients occur in the absence of adrenal metastases.

Our patient had characteristic cytological features of primary ACC of the lung which correlated well with CT, bronchoscopic, and histological findings. Our case was rare in the sense that it was a case of pulmonary ACC that had metastasized to the liver and kidneys at the time of initial presentation. Moreover, the patient was only 28 years old and had a predominantly cribriform pattern of ACC. Renal metastases were bilateral and occurred in absence of adrenal metastases.

References

Figure 3. Abdominal CT scan showing multiple metastatic lesions in both kidneys and liver.
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