

to polygonal and vesicular nucleus, small nucleolus, and scant to moderate eosinophilic cytoplasm, while the basal cells were relatively monotonous with small, round to angulated hyperchromatic nuclei and scant cytoplasm. Scattered mitotic figures were seen (Figures 1a and 1b). Lymphovascular and perineural invasion were not seen. No lepidic pattern of growth was identified.

A battery of immunostains performed showed a CD117+ (luminal)/CK5/6 (basal)/p63+ (basal)/DOG1-/TTF1-/Napsin A- phenotype (Figures 2a to 2c). The Ki-67 proliferation index was 20% (Figure 2d). Constellation of the histomorphologic features and immunohistochemical profile supported a diagnosis of adenoid cystic carcinoma (ACC). In the absence of a salivary gland lesion or any other mass elsewhere in the body, in spite of bilaterality of the masses, a diagnosis of primary pulmonary ACC was rendered.

## **PATHOLOGY PROFILE FINAL DIAGNOSIS**

### **PRIMARY PULMONARY ADENOID CYSTIC CARCINOMA**

#### **DISCUSSION:**

Primary pulmonary ACC is a relatively rare malignant tumor accounting for about 0.04-0.2% of all lung cancers. It is a slow growing tumor with a prolonged and indolent clinical course. They are known to arise from the tracheobronchial glands, so are more common in the central bronchi. Therefore, pneumocyte-associated markers such as TTF1 and Napsin A are typically negative. ACC arising in the peripheral lung must therefore be distinguished from a metastatic tumor.

As the morphology and immunoprofile of pulmonary and extrapulmonary ACC are similar, and there is no specific imaging profile to distinguish these two entities, anatomic location of the primary mass lesion and relevant clinical information are of paramount importance to render a correct diagnosis. ACC may occasionally mimic a carcinoid, cribriform variant of pulmonary adenocarcinoma, mucinous adenocarcinoma, and polymorphous low-grade adenocarcinomas (PLGA), on morphology. This is further compounded at the metastatic location. In such scenarios, immunohistochemistry can be a useful adjunct to the morphology to enhance the accuracy of histologic diagnosis. Of note, TTF-1, a highly sensitive and specific marker for diagnosis of primary lung cancer; however, the TTF-1