Adenoid Cystic Carcinoma: One Patient, Four Tumors, and Twenty Years

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Introduction

- Adenoid Cystic Carcinoma (AdCC) is a rare biphasic neoplasm comprised of epithelial and myoepithelial cells. Although AdCC is most often associated with salivary glands, it also arises in other locations such as breast and lung. AdCC arising in the breast accounts for 0.1-3.5% of all primary breast tumors, while lung AdCC, is even more infrequent, accounting for <1% of all primary lung tumors. This tumor is associated with genomic alterations of the *MYB* oncogene. Despite its triple negative phenotype, this tumor generally has a favorable prognosis with rare instances of metastasis; radical surgical excision is usually curative. Our case follows the course of a patient with an atypical presentation of AdCC.

Methods

- We received a consult from an outside provider (OSP) of a colon biopsy from a 75-year-old female. The endoscopist described a “polyp” in the colon, and the only information provided was “history of breast cancer.” The histomorphology and immunohistochemical profile were suggestive of metastatic disease, rather than primary colonic neoplasia. Additional history from the OSP clarified the breast cancer history as AdCC status post resection in 2005. Continued investigation revealed the patient presented with both lung and breast masses in 2023. We received both cases for review and they demonstrated similar morphology to the colon biopsy. Over the span of two decades, this patient presented with AdCC in three separate locations.

Pathologic Examination

- The initial colon biopsy revealed infiltrating malignant cells within the submucosa, arranged in cords and nests with cribriform architecture. No surface epithelial dysplasia was identified.

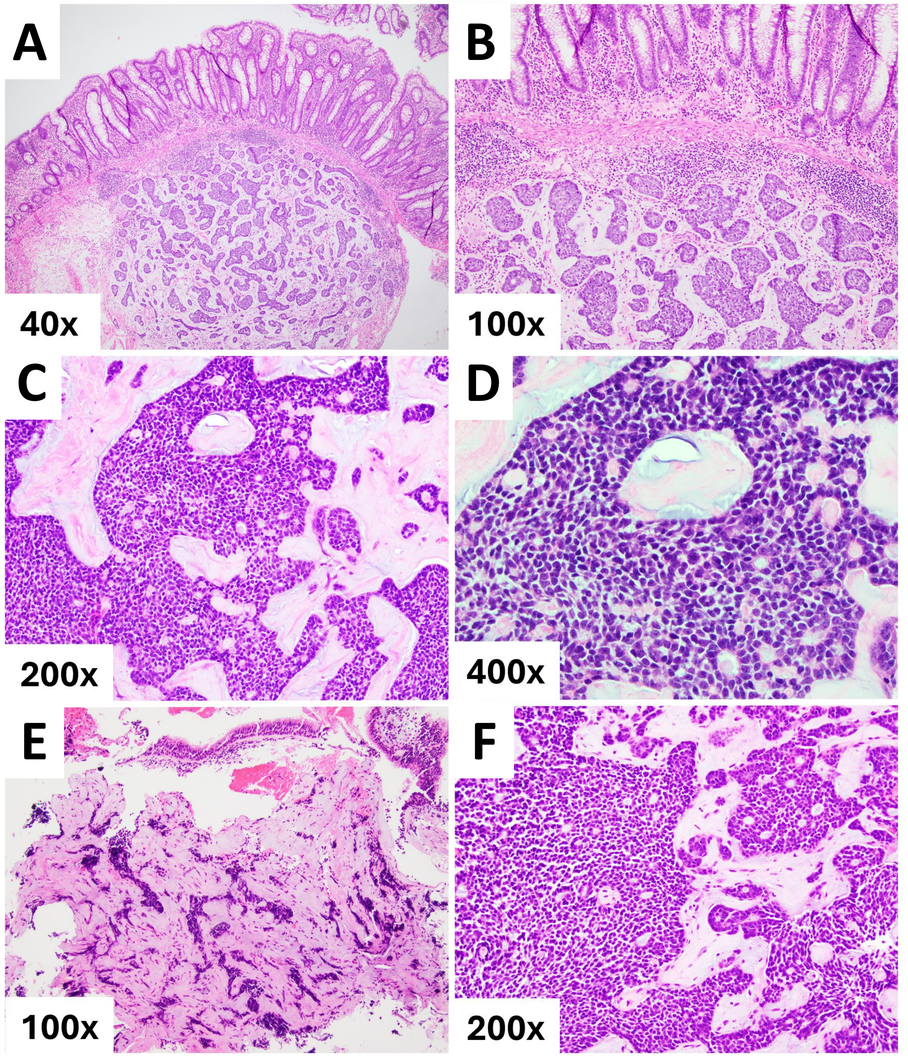

Figure 1: (A-B) Colon biopsy highlighting neoplastic cells confined to the submucosa, demonstrating cribriform architecture; (C-D) Breast resection showing glands lined by ductal epithelial cells and pseudolumens containing eosinophilic basement membrane material; (E-F) Lung biopsy exhibiting tumor cells embedded in extracellular matrix, in addition to cribriform areas.

Pathologic Examination

- Overall, these findings were consistent with metastatic disease; however, immunohistochemistry was nonspecific for a site of origin. The right breast lumpectomy and endobronchial biopsy demonstrated a similar morphology, and immunohistochemistry (IHC) demonstrated the presence of true glands lined by ductal epithelial cells (CD117+, CK7+) and pseudolumens lined by myoepithelial cells (p63+, SOX10+), confirming the biphasic nature of the tumor. MYB IHC further supported the diagnosis of AdCC.

Conclusions

- Although the piecemeal presentation of this case made it more difficult to draw appropriate conclusions, ultimately a complete clinical history, in conjunction with histologic evaluation of multiple cases allowed us to render the correct diagnosis. While recurrence decades after treatment is not unheard of, the subsequent metastatic disease involving the lung and colon represents a rare occurrence, as these tumors tend to be more indolent. Our case demonstrates the importance of a complete history in conjunction with thorough histologic examination when confronted with a challenging case. These factors are paramount to ultimately provide the correct diagnosis and appropriately guide patient care.

References and Acknowledgements

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2. WHO Classification of Tumours Editorial Board. Thoracic Tumours. 5th ed. Lyon, France: International Agency for Research on Cancer; 2021.

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