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Hepatoid Adenocarcinoma ofLung: A Rare and Challenging Tumor

Hepatoid adenocarcinoma (HAC) is an extremely rare extra-hepatic malignant tumor having hepatic features on pathology. Stomach is the most common site, and hepatoid adenocarcinoma of lung (HAL) accounts for 2.3 – 5% of all HACs. Less than 100 cases of HAL have been reported worldwide since its concept was put forward in 1990.

A 63-year-old female presented to the ED with back pain. Initial evaluation revealed anemia (hemoglobin 6.7), hypercalcemia (12.1 mg/dl), GFR 54 mL/min. X-ray thoracic spine and chest showed two vertebral compression deformities and lytic lesions on multiple ribs. Initially, multiple myeloma was considered likely but further evaluation only showed IgA lambda monoclonal gammopathy of 0.2g/dl. MRI thoracic spine done for further characterization of the vertebral deformities showed multiple abnormally enhancing lesions concerning for metastatic disease. CT chest/abdomen/pelvis revealed left perihilar lung mass measuring 5.1cm, numerous mediastinal and supraclavicular lymph nodes, and diffuse osseous and adrenal metastases. She then underwent bone marrow showing <10 % plasma cells, favoring MGUS, and evidence of metastatic carcinoma. Concurrently done supraclavicular lymph node biopsy exhibited evidence of poorly differentiated carcinoma. Immunohistochemistry was positive for Hep-par-1 suggesting hepatic origin but was negative for arginase, glypican-3, and WT1 arguing against primary hepatocellular malignancy. With large lung mass, lack of liver lesions, presence of adrenal metastases, and immunohistochemistry results, diagnosis of HAL was made. Serum AFP level however was normal. With advanced disease, the patient declined treatment and was enrolled in hospice.

HAL is extremely rare, and tumor characteristics, prognosis, treatment strategies have been proposed based only on a few retrospective studies. It is diagnosed with presence of both adenoid and hepatocyte-like differentiation either on pathology or immunohistochemistry. Its diagnosis can be challenging based on morphology alone, especially if poorly differentiated as in our patient. So, immunohistochemistry is the cornerstone for accurate diagnosis. Elevated serum AFP is seen in a majority of cases and is a poor prognostic indicator, but as in our case, it is not required for diagnosis. It is a rapidly growing carcinoma with a poor prognosis, so early intervention is essential to reduce the tumor burden and complications of enlarging lung mass.