Metastatic epithelioid angiosarcoma with bone marrow involvement

A previously healthy 49-year-old woman presented with a 2-month history of unintentional weight loss, early satiety, nausea, and vomiting. Whole-body computerized tomography scan revealed hepatosplenomegaly with enhancing lesions in the liver, spleen, lumbar vertebrae, and right iliac bone. A liver biopsy sample was initially interpreted as a hemangioma. Complete blood count revealed marked anemia with hemoglobin 7.8 g/dL, a white blood cell count of $25.4 \times 10^9/L$, and a platelet count of $135 \times 10^9/L$. Peripheral smear demonstrated a leukoerythroblastic picture. A left iliac bone marrow biopsy sample revealed a hypercellular marrow with osterosclerotic changes and focal involvement by neoplastic cells (panel A). Large sheets of abnormal, round to polygonal epithelioid cells, along with focal areas of irregularly anastomosing malignant vessel formation, were noted (panels A-B; arrow in panel A indicates sarcoma cells). By immunohistochemistry (IHC) studies, these tumor cells were strongly positive for CD31, CD34, Friend leukemia virus integration 1 (FLI1), and erythroblast transformation–specific related gene (ERG) stains (panels C-F) and negative for cytokeratin, CAM5.2, EMA, MOC31, CD3, CD20, CD30, CD138, MPO, S-100, BRST-2, HMB45, actin, desmin, CD117, factor 8, podoplanin, PAX-8, ALK-1, and DOG1 stains.

The patient was diagnosed with metastatic epithelioid angiosarcoma. Involvement of the bone marrow is an extremely rare presentation of this aggressive malignancy. Our case also demonstrates the utilization of the recently described, highly specific IHC markers such as ERG and FLI1 in identifying such rare neoplasms.
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