Pathologic Quiz Case

Elham Abboud, MD; Elliot Weisenberg, MD; Sanobar Khan, MD; Douglas P. Rhone, MD; Chicago, Ill

A 65-year-old white woman presented to her primary care physician with the complaint of right upper quadrant pain. Her medical history was remarkable for hypertension, diabetes mellitus, and asthma. She was evaluated for cholelithiasis, and gallstones were detected. A 6.0-cm right adrenal mass was detected by computed tomographic scan (Figure 1). Evaluation of the chest, abdomen, and pelvis, including computed tomographic scanning and magnetic resonance imaging revealed no other lesions. Determination of urinary cortisol and vanillylmandelic acid levels showed no evidence of a functioning tumor. Cholecystectomy and excision of the right adrenal mass were performed.

The gallbladder showed chronic cholecystitis and cholelithiasis. The right adrenalectomy specimen consisted of an 8.0 × 7.5 × 5.5-cm ovoid mass with a focally hemorrhagic surface. The hemorrhagic oval mass formed nearly 90% of the specimen, was fairly well circumscribed, and was surrounded by a rim of flattened adrenal tissue. Cut section revealed a variegated cut surface (Figure 2). Histopathologic examination revealed ovoid nests, papillary fronds, and solid areas of tumor cells, which on occasion formed vascular channels containing red blood cells. The tumor cells were large, round to oval, and markedly atypical with moderately abundant amphophilic cytoplasm and poorly developed cell borders. The nuclei were vesicular, round to oval, and contained prominent eosinophilic nucleoli. Brisk mitotic activity was present, and atypical mitotic figures were identified. Occasional intracytoplasmic lumina contained red blood cells (Figure 3). The tumor infiltrated residual adrenal cortical tissue in small clusters and as single cells. The capsule of the adrenal capsule appeared intact. Immunohistochemical studies showed strong staining for factor VIII–related antigen (Figure 4), vimentin, and focal positivity for pancytokeratin. Staining for neuron specific enolase and synaptophysin were not detected.

What is your diagnosis?
Pathologic Diagnosis: Epithelioid Angiosarcoma of the Adrenal Gland

Angiosarcomas are rare tumors estimated to represent fewer than 1% of all sarcomas. Skin and soft tissue are the most common sites; however, about one quarter of cases are found in other locations, such as breast, liver, bone, and spleen. Angiosarcoma of the adrenal gland is extraordinarily rare. Fewer than 20 cases have been reported in the English literature. In all the reports in which histopathologic characteristics are described, the tumors have exhibited epithelioid features typical of deeply situated tumors. The finding of reactivity for cytokeratin is typical of epithelioid morphology and is believed to represent either aberrant or "atavistic" expression.

The first case was described in 1988 by Kareti et al in 1988. The largest study of epithelioid angiosarcoma of the adrenal gland was done at the Armed Forces Institute of Pathology by Wenig et al in which 9 cases, including the first case, were described. Three patients died with metastatic disease, 1 patient died in the immediate postoperative period, 1 case was found incidentally at autopsy, and 3 patients were alive without evidence of disease from 6 to 13 years after initial resection. The patient reported by Kareti et al died 4 years after adrenalectomy and wide en bloc resection without evidence of disease. Our patient received only surgery and is alive and well without evidence of recurrent tumor 1 year after surgery.

The other reports are single cases. One patient died in the immediate postoperative period, 1 died with widely metastatic disease, and 2 were apparently cured, 1 with surgery and radiation therapy and 1 with surgery alone. The patients have ranged in age from 41 to 85 years old with a mean of about 60 years. Tumors have been reported in the English literature in 6 women and 8 men, including the present case. In other sites, angiosarcomas have been associated with radiation exposure, chronic lymphedema, and exposure to vinyl chloride. In the adrenal gland 1 patient suffered from chronic arsenic intoxication and another had accompanying mesenteric fibrosis. One patient came to medical attention because of a paraneoplastic syndrome consisting of cough, malaise, fever, anemia, and elevated alkaline phosphatase, which resolved after resection of the tumor.

Epithelioid angiosarcoma of the adrenal gland may mimic much more common primary and secondary tumors and, in view of cytokeratin positivity, especially metastatic carcinoma. Despite its rarity, knowledge of its existence is important as its pathobiologic characteristics may differ markedly from other primary and metastatic adrenal neoplasms. Because of the infrequency of this entity optimal therapy other than surgical eradication is difficult to determine. Several patients, including ours, have done well following complete surgical resection of the tumor. Some cases may have been detected at an early enough stage to enable surgical cure. In view of the aggressive nature of angiosarcoma in all sites, adjuvant therapy appears justified for patients in whom complete surgical extirpation cannot be ensured.

References