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Angiosarcoma of the Adrenal Gland Treated Using a Multimodal Approach

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ABSTRACT

Angiosarcoma of the adrenal gland is an extremely rare malignancy. We report a case of a 59-year-old female who presented with abdominal pain and profound anemia. A 7 cm enhancing, lipid-poor adrenal mass with calcifications that extended posterior to the vena cava was identified on imaging. Patient underwent right adrenalectomy with retroperitoneal lymph node dissection. Pathology demonstrated angiosarcoma of the adrenal gland. Consolidative treatment included adjuvant radiation and chemotherapy. Patient remains disease free 1.5 years following treatment. Prior reported literature on the diagnosis and management of adrenal angiosarcoma is reviewed.

Introduction

Angiosarcoma is a malignant endothelial-derived, soft-tissue sarcoma that arises from vascular or lymphatic origin.[1] The majority is thought to arise spontaneously; however, malignant transformation from within benign vascular lesions has also been reported.[1] The most common locations for this tumor are the head and neck (27.0%), breast (19.7%), and the extremities (15.3%).[1]

Angiosarcoma of the adrenal gland is an exceedingly rare malignancy with less than 50 reported cases in the literature to date.[2–27] The patient described herein presented initially with abdominal pain and severe anemia, was found to have a right adrenal mass upon a diagnostic workup, and then received multimodal therapy in the form of surgery, adjuvant chemotherapy and radiation.

Case presentation

A 59-year-old female with no history of prior malignant disease presented with a chief complaint of abdominal pain and anemia. Her hemoglobin (HGB) 10 weeks prior to presentation was 7.8 gm/dL, and leukocytosis with a white blood cell count of ~18 k/mm^3

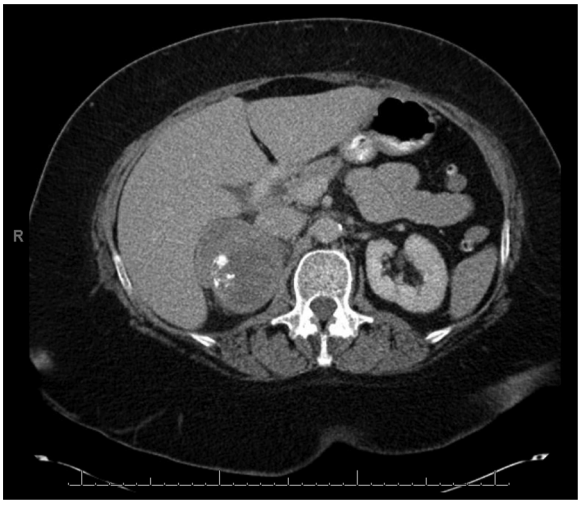

Figure 1. Enhancing, lipid-poor mass with coarse areas of calcification measuring 7.0 × 6.7 cm in the right adrenal gland. Note relationship of the mass to the vena cava.

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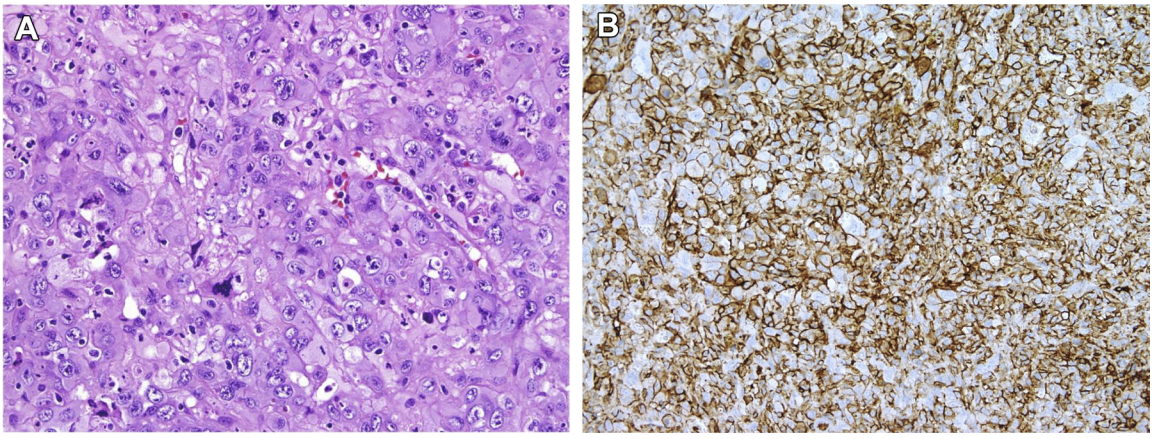

Figure 2. (A) Angiosarcoma of the adrenal gland. High power magnification demonstrates a sheet of large pleomorphic, round epithelioid cells with eosinophilic cytoplasm and large hyperchromatic nuclei with prominent nucleoli. Scattered intracytoplasmic lumina contain erythrocytes. Numerous mitotic figures are present (H & E, 40× magnification). (B) CD31 immunohistochemical stain. Diffuse intense membranous and cytoplasmic tumor cell staining with this vascular marker is apparent. (40× magnification).

was also present. An abdominal ultrasound revealed a 7 cm (cm) retroperitoneal mass arising from the right adrenal gland. To better characterize the mass, a computerized tomography (CT) scan was performed, which demonstrated a 7 cm enhancing, lipid-poor adrenal mass with coarse areas of calcification concerning for a primary adrenal cortical carcinoma (Fig. 1). Serum aldosterone, renin, testosterone, DHEA-S, estradiol, androstenedione, 17-OH progesterone, metanephrine, and normetanephrine levels were within normal limits, and serum cortisol was slightly elevated at 7.6 μg/dL upon low dose dexamethasone suppression testing. The patient underwent an open right adrenalectomy with lymph node dissection. Pathology demonstrated an 11.5 × 8.5 × 6.0 cm adrenal mass with invasion into the peri-adrenal fat. The tumor was within <0.01 cm margin of resection where it was densely adherent to the vena cava. Paracaval lymph nodes were negative for malignancy. Immunohistochemistry was positive for CD31, chromogranin, vimentin, and epithelial membrane antigen, and negative for calretinin, synaptophysin, TTF-1, GATA3, A103, AE1/AE3, CD5, and inhibin (Fig. 2). As such, these findings were consistent with angiosarcoma.

After a multidisciplinary discussion, patient underwent 5 of 6 planned cycles of adjuvant chemotherapy with doxorubicin, ifosfamide, and mesna (AIM) followed by external beam radiotherapy (XRT) in 25 fractions for a total of 50 Gray (Gy). Her anemia improved upon resection of the mass, with a HGB increase to 11.5 gm/dL 7 weeks after surgery and resolution of leukocytosis (WBC count of 6.5 k/mm^3 on postoperative day 20). Morning cortisol on postoperative day 4 was normal at 14.6 μg/dL. At 18 months following completion of treatment, the patient remains without radiographic evidence of disease.

Discussion

We report adrenalectomy followed by adjuvant XRT and chemotherapy for localized adrenal angiosarcoma. At eighteen months, the patient remains disease-free.

Angiosarcoma of the adrenal gland was first described in 1988 by Kareti and colleagues.[2] Since then, an additional 40 cases have been reported in the literature with a median age at presentation of 60 years (range 34–85 years).[3–27] Patients present in a variety of ways including flank or abdominal pain, palpable mass, fatigue, weight loss, fever, or clinically asymptomatic with a mass detected on incidental imaging. Table 1 summarizes the age, sex, type of presentation, treatment, and follow-up for previously reported cases.[2–27] Patients who present with systemic symptoms tend to have worse outcomes than those with localized symptomatology.

Given the rarity of the disease, there is no consensus guideline on the best treatment algorithm for adrenal angiosarcomas. Adrenalectomy serves both diagnostic and therapeutic purposes. For patients with localized disease, the median survival is 7 months,[1] while one study reported a 2-year overall survival of 21%.[28] Adjuvant radiation therapy has been previously reported and was elected for our patient due to proximity of the surgical margin at the posterior vena cava. Previously reported multimodal treatment approaches have included doxorubicin-based chemotherapeutic regimens and adjuvant radiation therapy (XRT).[28] These data suggested risk reduction of systemic progression and improved 2-year survival in patients with visceral angiosarcoma who undergo multimodal treatment consisting of extirpation, adjuvant chemotherapy, and in some cases radiation.[28,29] Furthermore, a meta-analysis assessing the use of adjuvant doxorubicin-based chemotherapy for localized resectable soft-tissue sarcoma demonstrated a decreased risk of local, distant, and overall recurrence, and improved survival.[30] For our patient, restaging scans after completion of systemic therapy demonstrated no signs of local recurrence or distant metastatic disease. Adjuvant XRT has been shown to potentially improve disease-free survival in patients with angiosarcoma.[31] Thus, given the focally close margin at the posterior vena cava and no evidence of progression, 50 Gy of radiation was delivered to the operative bed using a modulated technique respecting normal tissue thresholds of the adjacent bowel and kidneys. Treatment was tolerated without event.