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Case Report

Primary perirenal angiosarcoma: A rare presentation of a perirenal mass

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Introduction

Angiosarcoma is a rare soft tissue tumor that accounts for only 2% of sarcomas [1]. It originates from vascular and lymphatic endothelium and most commonly arises in the head, neck, and breast [2]. In this report, we present an unusual presentation of a primary perirenal angiosarcoma.

Case report

A 65-year-old male presented to his primary care physician with a 6-month history of left flank pain and an unintentional 20-pound weight loss. His prior medical history was notable for a 30-pack year smoking history, but was otherwise unremarkable. He was anemic with a hematocrit of 30.5 (reference range 41.0%–53.0%), but his hemoglobin was 13 g/dL (reference range: 13.5–17.5 g/dL) and his serum creatinine was 0.8 mg/dL (reference range: 0.60-1.50 mg/dL), both normal. He denied hematuria, headache or bone pain. Physical exam revealed a palpable left flank mass. A contrast material-enhanced computed tomography (CT) scan of the abdomen and pelvis was obtained and demonstrated a 7.2 × 4.0 cm, enhancing soft tissue mass arising from the left perirenal space (Fig. 1). The right kidney was atrophic and there was no evidence of lymphadenopathy or invasion of the left renal vein. A CT-guided percutaneous biopsy of the soft tissue mass established a diagnosis of perirenal angiosarcoma. Histochemical analysis revealed tumor cells were diffusely positive for CD31 and FLI-1 but negative for MNF116 and D2-40. Aki-67 stain revealed a proliferative index of approximately 40%.

Because of the history of an atrophic right kidney and the desire to avoid a left nephrectomy and the subsequent need for long-term hemodialysis, the patient was treated with Taxotere followed by proton beam therapy (18 Gy). One month later (Fig. 2), he underwent left flank exploration with resection of left perinephric tumor and partial left nephrectomy. Surgical pathology revealed extensive tumor necrosis and giant cell reaction and no viable tumor cells.

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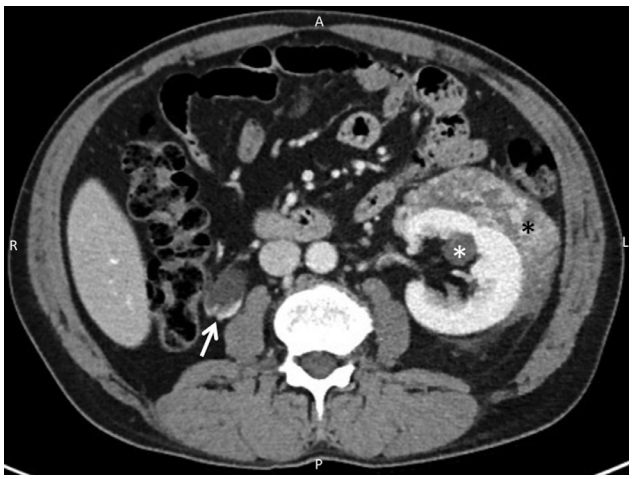

Fig. 1 – Axial contrast-enhanced CT scan of the abdomen that demonstrates an enhancing left perirenal soft tissue mass (black asterisk) subsequently biopsy proven to represent perirenal angiosacoma. White arrow indicates atrophic right kidney (white arrow). White asterisk indicates a benign cyst.

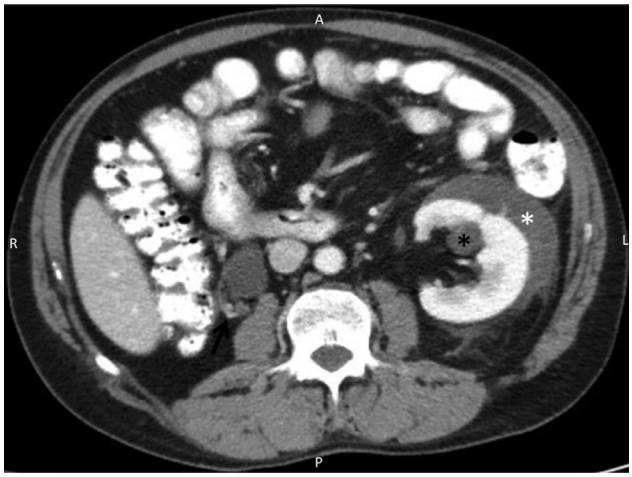

Fig. 2 – Axial contrast enhanced CT scan of the abdomen after chemoradiation of left perirenal angiosarcoma (white asterisk). The mass has diminished slightly in size and no longer shows enhancement. The patient subsequently underwent resection of the mass and the pathology shows no viable tumor. Black asterisk indicates a benign cyst.

Discussion

Perirenal angiosarcoma is an exceedingly rare malignant neoplasm [3]. Most reported cases of renal angiosarcoma represent metastasis from other sites [4]. Very few cases of renal angiosarcoma have been reported in the literature [5]. Primary renal angiosarcomas often occur in 6th decade of life with predilection for men [6,7]. Risk factors for renal angiosarcoma have not yet been identified; however, exposure to radiation and vinyl chloride are known risk factors for soft tissue and hepatic angiosarcomas [8,9].

Early diagnosis is challenging because clinical symptoms often develop when tumor has metastasized to other organs [3]. Radiologic imaging alone cannot provide sufficient information for diagnosis. CT findings widely vary for this condition, they can present as heterogenous mass, as in our case, or a hypodense mass with areas of enhancement [6,10]. Diagnosis is only possible after immunohistochemical staining. The most commonly used markers are CD-31, CD-34 which are endothelial adhesion molecule and a human hematopoietic progenitor cell antigen [11,12].

Because of their rarity, there is no standard therapy for these tumors. While nephrectomy is often performed in most reported cases [6], this case demonstrates the clinical effectiveness of neoadjuvant chemoradiation prior to renal preserving resection of the mass.

REFERENCES

[1] Rouhani P, Fletcher CD, Devesa SS, Toro JR. Cutaneous soft tissue sarcoma incidence patterns in the U.S.: an analysis of 12,114 cases. Cancer 2008;113:616–27.
[2] Ray-Coquard I, Italiano A, Bompas E, et al. Sorafenib for patients with advanced angiosarcoma: a phase II Trial from the French Sarcoma Group (GSF/GETO). Oncologist 2012;17:260–6.
[3] Yoshida K, Ito F, Nakazawa H, Maeda Y, Tomoe H, Alba M. A case of primary renal angiosarcoma. Rare Tumors 2009;28:12–28.
[4] Lee C, Park K, Nah D, Won K. Bilateral spontaneous pneumothorax during cytotoxic chemotherapy for angiosarcoma of the scalp: a case report. J Korean Med Sci 2003;18:277–80.
[5] Omiyale AO, Carton J. Clinical and Pathological features of primary angiosarcoma of the kidney. Curr Urol Rep 2018;19:4. doi:10.111/tbj.13006.
[6] Leggio L, Addolorato G, Abenavoli L, et al. Primary renal angiosarcoma: a rare malignancy. A case report and review of the literature. Urol Oncol 2006;24:307–12.
[7] Brown JG, Folpe AL, Rao P, et al. Primary vascular tumors and tumor-like lesions of the kidney: a clinicopathologic analysis of 25 cases. Am J Surg Pathol 2010;34:942–9.
[8] Johnson V, Gaertner E, Crothers B. Fine-needle aspiration of renal angiosarcoma. Arch Pathol Lab Med 2002;126:478–80.
[9] Hiratsuka Y, Nishimura H, Kajiwara I, Matsuoka H, Kawamura K. Renal angiosarcoma: a cse report. Int J Urol 1997;4:90–3.
[10] Peters HJ, Nuri M, Münzenmaier R. Hemangioendothelioma of the kidney: A case report and review of the literature. J Urol 1974;112:723–6.
[11] Leggio L, Addolorato G, Abenavoli L, et al. Primary renal angiosarcoma: a rare malignancy. a case report and review of the literature. Urol Oncol 2006;24:307–12.
[12] Sabharval S, John NT, Kumar RM, Kekre NS. Primary renal angiosarcoma. Indian J Urol 2013;29:145–7.