RESIDENT'S CORNER

Epithelioid angiomyolipoma metastasis to the rectus abdominis

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A 37-year-old female presented with abdominal pain. An abdominal computed tomography scan demonstrated a 10 cm x 13 cm left renal mass. An open adrenal-sparing radical nephrectomy was performed. The pathological diagnosis was epithelioid angiomyolipoma. Five-year surveillance did not demonstrate recurrence of disease. However, a 1.8 cm x 2.5 cm mass on the rectus abdominis muscle was identified after 5 years. Biopsy of the mass demonstrated histologic findings consistent with the primary tumor. Herein, we report a case of metastatic renal epithelioid angiomyolipoma to the rectus abdominis muscle more than 5 years after resection of primary renal tumor.

Key Words: renal epithelioid angiomyolipoma, perivascular epithelioid cell tumor, PEComa, metastasis, rectus abdominis

Introduction

Epithelioid angiomyolipomas also known as perivascular epithelioid cell tumors (PEComa) of the kidney are a rare tumor histologically similar to renal angiomyolipoma, characterized by proliferation of predominantly epithelioid cells.[1] They account for less than 10% of renal tumors with incidence of 0.3% in the general population.[2] These tumors can exhibit aggressive behavior, with approximately 50% of reported cases demonstrating disease progression.[1] Some of the more common metastatic sites that have been reported are the liver, lymph nodes, lung, and bone. Metastasis to the ileum has also been reported.[3] According to World Health Organization, approximately one-third of patients with epithelioid angiomyolipoma have metastasis to lymph nodes, liver, lungs, or spine.[4] Herein, we report a case of renal epithelioid angiomyolipoma that metastasized to the rectus abdominis muscle 5 years after resection of primary renal tumor.

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Case report

A 37-year-old obese female with no other significant past medical history presented with a chief complaint of abdominal pain. On computed tomography (CT) with intravenous contrast, she was found to have a 10 cm x 13 cm left upper pole enhancing renal mass, Figure 1. The patient was referred to the urology service and she underwent an adrenal-sparing left radical nephrectomy via flank incision in the standard fashion. Intraoperative findings were notable for substantial perirenal neovascularity. Estimated blood loss was 700 mL. There were no intraoperative complications. The patient had an unremarkable postoperative course and was discharged to home on postoperative day 3.

Initial pathologic analysis was reported as consistent with renal cell carcinoma, favoring chromophobe type with lymphovascular invasion and focal central necrosis. On immunohistochemistry, the tumor cells were positive for c-KIT, but not for CK7, CD10, and racemase. Given the discordance, the specimen was sent to the Cleveland Clinic for expert opinion, where the diagnosis of perivascular epithelioid cell tumor was made. The tumor was 13 cm x 12 cm x 8.5 cm in diameter with significant perinephric neovascularization. All surgical and vein margins were negative for tumor. The mass was 2.5 cm from the vascular margin of resection. The mass grossly appeared to be confined within the kidney capsule. Microscopically, the tumor

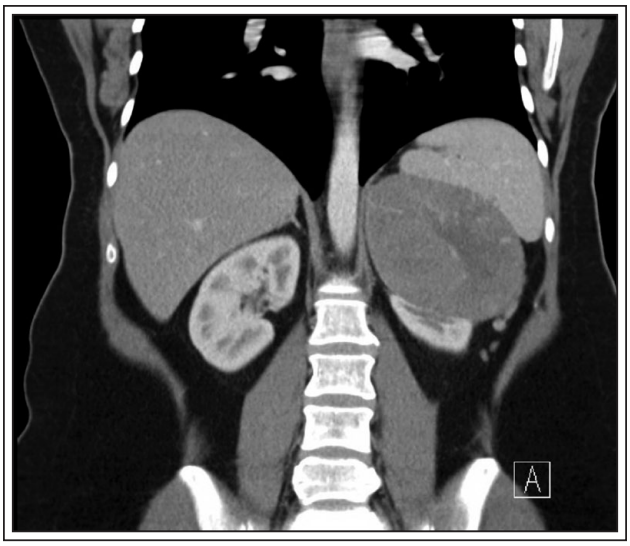

Figure 1. CT abdomen/pelvis with contrast demonstrating a 13 cm left upper pole enhancing renal mass.

demonstrated focal central necrosis and some cellular atypia with focal angiolymphatic invasion in a single site. On immunohistochemistry, the neoplastic cells stained positive for CD117, Melan-A, and HMB45. Cells stained negative for CK7, CA9, smooth muscle actin, desmin, HHF35 actin, epithelial membrane antigen, S100, CAM 5.2, and AE1/3.

Due to the potentially malignant nature of these tumors, the patient underwent surveillance CT of the chest, abdomen, and pelvis. On the initial scans, there was no evidence of local recurrence or metastatic disease. However, approximately 18 months following radical nephrectomy, CT of the abdomen demonstrated two new para-aortic masses measuring 3.1 cm x 3.6 cm and 1.9 x 2.3 cm which were concerning for regional lymphadenopathy, Figure 2.

A PET scan was performed subsequently which failed to demonstrate any FDG avidity. A percutaneous biopsy of the presumed lymphatic recurrence was performed but was nondiagnostic, demonstrating fragments of fibro-connective tissue with focal histiocytic infiltrate, skeletal muscle, and necrotic material. Resection of the retroperitoneal node was considered, but deferred by the patient and she continued to undergo surveillance. Interestingly, over the next 2 years, subsequent abdominal CT demonstrated that the lymphadenopathy progressively decreased in size, such that by 31 months following diagnosis, there was no evidence of regional adenopathy in the absence of definitive intervention. However, on subsequent CT, the mass was again measured at 1.4 cm in diameter. A regional lymphadenectomy was again considered, however, the patient elected to continue surveillance.

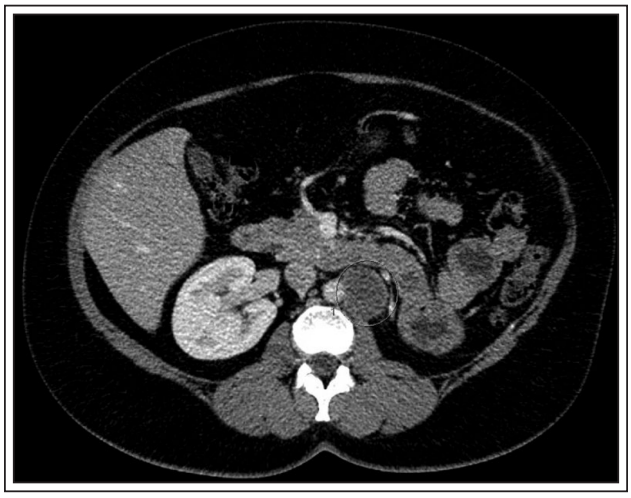

Figure 2. CT abdomen/pelvis with contrast demonstrating a 3.1 cm x 3.6 cm left para-aortic lymphadenopathy.

On subsequent CT of the abdomen performed 5.5 years following nephrectomy, the para-aortic nodes remained stable, however a soft tissue nodule abutting the left rectus abdominis muscle measuring 1.8 cm x 2.5 cm was identified, Figure 3. The patient underwent ultrasound-guided biopsy of the nodule, which demonstrated tumor cells sharing morphologic features of original tumor with focal necrosis with a similar immunohistochemical profile positive for melan-A, HMB-45, CD117, but not smooth muscle actin, epithelial membrane antigen, and desmin.

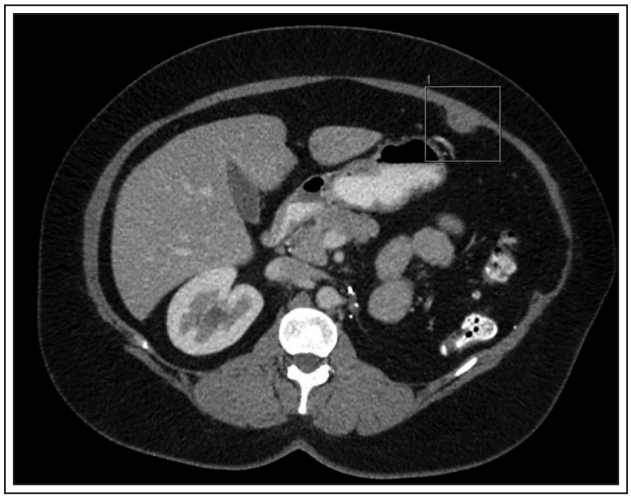

Figure 3. CT abdomen/pelvis with contrast demonstrating a new soft tissue nodule abutting the left rectus abdominis muscle measuring 1.8 cm x 2.5 cm.

This was deemed consistent with metastatic perivascular epithelioid tumor. She was offered excision, but again deferred. On subsequent CT scans, the mass increased in size from 2.9 cm x 2.3 cm. Over the next 6 months, it increased in size to 2.7 cm x 3.5 cm in diameter. Given the growth kinetics of the lesion, the patient agreed to undergo surgical excision of the subrectus mass and is awaiting the upcoming surgery.

Discussion

Renal epithelioid angiomyolipomas represent a rare variant of renal neoplasm. These tumors are classified as part of a family of mesenchymal tumors called perivascular epithelioid cell tumors (PEComa). PEC tumors are characterized by perivascular clear cell and epithelioid features that co-express melanocytic and muscle markers. It is considered a benign neoplasm and consists of thick-walled poorly organized blood vessels, smooth muscle, and varying levels of mature adipose tissue.[5] The tumor strongly expresses estrogen, progesterone, and androgen receptors. They are predominantly found in females after puberty, suggesting a potential hormonal influence.[6]

Renal epithelioid angiomyolipomas are potentially malignant and can be locally aggressive or metastasize. Liver, lung and bone are more commonly reported sites of metastasis. After reviewing 234 cases of perivascular epithelioid cell tumors, Bleeker et al reported an association between tumor size > 5 cm and high mitotic rate (> 1/50HPF) and increased risk of recurrence.[7]

The patient in this case report had no definitive evidence of metastasis for nearly 5 years of surveillance following initial extirpation of the renal tumor. CT scan at 5 years demonstrated a soft tissue nodule abutting the rectus abdominis muscle. The nodule had been biopsied and is demonstrated to be a perivascular epithelioid tumor, which shares morphologic and immunohistochemical features with the renal mass resected 5 years prior. To the authors' knowledge, this is the first report of a renal PEComa metastasis to muscle tissue. As the surgical approach was performed via a left flank and the rectus abdominis muscle was not violated, seeding of the muscle is unlikely.

Metastasis after nephrectomy for a PEComa is not a frequent occurrence, but prior studies have presented cases with metastasis occurring 12 months postoperatively.[8] One case study presented a case of renal PEComa with metastasis to the lungs and ileum 5 years after surgical excision.[3] To our knowledge, the current case was the first reported case of metastatic PEComa to the abdominal wall. This presentation is particularly unique given clean margins shown on pathology and the significant delay between nephrectomy and recurrence, and the location of metastasis. □

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