Successful management of renal leiomyosarcoma using radiofrequency ablation: Case report and brief review of literature

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Received: March 12, 2014 Accepted: July 10, 2014 Online Published: August 7, 2014

DOI: 10.5430/jst.v4n3p10 URL: http://dx.doi.org/10.5430/jst.v4n3p10

Abstract
Metachronous Leiomyosarcoma are a rare entity in literature. Renal Leiomyosarcoma (LMS) have an aggressive biological behavior with poor prognosis despite surgery. We report the use of a minimally invasive percutaneous Radiofrequency Ablation (RFA) in management of this tumor.

We report a case of 72 year old woman with a history of the inferior vena cava (IVC) LMS surgically treated 7 year ago and with metachronous/delayed presentation of subcutaneous LMS in scapular region and renal LMS, the later successfully treated with RFA. RFA seems to be an alternative procedure for patients with extended disease and comorbidities.

Key words
Leiomyosarcoma, Renal, Radiofrequency ablation

1 Introduction
Leiomyosarcoma (LMS) are soft tissue sarcomas arising from smooth muscle cells and are most commonly seen in the uterus, but can be seen anywhere in the body [1]. Independent of the organ of origin, the histologic appearance of LMS is similar, however clinical presentation varies depending of the organ involved. Diagnosis by imaging is challenging; and most diagnoses are made on post-surgical pathology. The overall prognosis of LMS is poor, surgery being the mainstay of treatment [1, 4].

We present a case of metachronous LMS involving the IVC, shoulder and kidney over different time intervals and management of kidney mass with Radiofrequency ablation (RFA).
2 Case report

A 72-year-old female with past history of hepatitis B, psoriasis and multiple surgeries including ovarian cystectomy, removal of an osteochondroma of the left hand and hysterectomy for fibroids presented 10 years ago with signs and symptoms of urinary tract infection.

A Chest and Abdominal computed tomography (CT) was performed which demonstrated a lesion which was thought to arise from the IVC at the level of insertion of the renal veins measuring 2.3 cm × 2 cm. She underwent complete surgical resection of the tumor. Pathology described as a high grade LMS with negative margins (Figure 3 a, b). Patient remained asymptomatic on a 7 year follow up with no recurrence on follow up imaging.

Thereafter upon clinical examination, a subcutaneous mass in the right scapular region was observed. An excisional biopsy revealed a high-grade LMS measuring 3.5 cm, confined to the subcutis. She underwent resection of the right scapular mass with reconstruction under general anesthesia. Additional imaging showed the interim development of a left lower pole renal mass measuring 2.4 cm × 2.8 cm. (Figure 1 A), and small pulmonary nodules which were not seen on the previous examination, which were never biopsied.

**Figure 1A.** CT scan performed revealed an enhancing mass (red arrows) measuring 2.4 cm × 2.8 cm arising from lower pole of the left kidney. **B.** CT scan performed 17 months later reveals an increase in size of the mass (red arrows) which now measures 3.4 cm × 3 cm. **C.** Coronal CT which shows the mass (red arrows). The differential diagnosis of this mass based on imaging was a renal cell carcinoma or a LMS involving the left kidney(due to previous history)

Seventeen-months later, the patient had follow-up imaging. CT of the abdomen revealed moderate increase in size of the exophytic left lower pole kidney lesion which now measured 3.4 cm × 3 cm, previously 2.4 cm × 2.2 cm (Figure 1B,C) .There were small left common iliac nodes. The patient was referred to the department of urology for management of the renal mass. In view of age, multiple prior surgeries and co-morbidities, she was deemed unfit for surgical resection and was referred for percutaneous CT scan guided RFA.

Two months later, she underwent CT scan guided RFA of the renal lesion (Figure 2 A, B, C). Patient was placed in prone position on the CT scan table and the procedure was performed under general anesthesia. Under continuous CT scan monitoring, in stages, first a biopsy was performed of the lesion and sent for frozen section and permanent slides (4 passes were performed) (Figure 2B).Biopsy performed concurrently confirmed the diagnosis of high-grade LMS.

Subsequently, radiofrequency ablation probe (LeVeen Needle Electrode 4.0, Boston Scientific Corp., Natick, MA) was advanced until the tip of the probe noted to be within the lesion. Once the position of the probe was confirmed, the probe was deployed to 4 cm and radiofrequency ablation was performed for according to the protocol. The probe was then pulled back 1 cm and the area retreated. Subsequent tract ablation was performed. CT scan was repeated which showed no procedure related complications. Patient tolerated the procedure well.
Figure 2. Radiofrequency Ablation procedure. A. Patient in prone position with grid placed on the back. B. Biopsy performed before ablation. C. Ablation performed using LeVeen Needle Electrode 4.0, Boston Scientific Corp., Natick, MA according to protocol.

Figure 3: (a, b). Microscopic appearance of inferior vena cava leiomyosarcoma; (a). Intersecting bundles of spindle cells with elongated and blunt-ended nuclei. (H&E, 20X) (b) Nuclear pleomorphic characteristics with mitosis (H&E, 60X); (c, d). Microscopic appearance of subcutaneous leiomyosarcoma from scapular region; (c). Intersecting bundles of spindle cells with elongated and blunt-ended nuclei. (H&E, 20X) (d) Nuclear pleomorphic characteristics with mitosis (H&E, 60X)

Follow Up imaging was performed with a CT scan at 1 month (Figure 4A), MR at 4, and 7 months (Figure 4B, C). No enhancement was seen within the ablated lesion to suggest residual or recurrent disease. Post-ablation changes were noted in the form of peri-nephric stranding. Clinically the patient has no complaints. She has gained some weight and appears to be in a good state of health.

3 Discussion

Leiomyosarcomas (LMSs) are a type of soft tissue sarcomas that arise from smooth muscles cells. Most (50%) of the tumors originate in the uterus with the other more common sites including retroperitoneum, skin, vessels, and bone. Extremely rare sites of primary locations include thyroid gland, gallbladder, base of tongue, liver, bronchus, kidney, and pancreas[1]. Morphologically these tumors are made of spindle cells arranged in fascicles with centrally located, elongated nuclei. Varying amounts of cellular atypia, necrosis and mitotic activity are seen in them. Over the years several muscle specific immunohistochemical markers, such as desmin and caldesmon, among others, have been recognized to aid in their
diagnosis\[1-3\]. Imaging can help in making a diagnosis of a LMS but it is not definitive. Large heterogeneous masses with irregular borders and necrosis can be seen both on CT and MRI. These features are nonspecific. Cross sectional imaging is more useful to evaluate the anatomical location and involvement of nearby vessels and organs. Diffusion-weighted MRI and PET using new radiotracers like 18F-FLT (18F-fluoro-3’-deoxy-3’-L: -fluorothymidine ) may help in differentiating benign from malignant tumors\[1, 4, 5\]. The uterine and extrauterine subtypes of LMS are histologically similar, but they have varying clinical behavior and prognosis depending on the organ involved. The overall prognosis of LMS is poor and the management of these sarcomas is challenging. Presently, the mainstay of treatment is surgery with clear, wide margins being the only curative approach with poor outcomes in cases where this is not possible. Recently radiation and chemotherapy are being used in a neoadjuvant/ adjuvant setting as well as in advanced/metastatic settings. Systemic chemotherapeutic agents and targeted therapy have become available to treat LMS and their use has increased in the past few years\[1, 6, 7\].

Figure 4. A. CT scan performed 1 month , B. MRI performed after 5 months, C. MRI performed after 13 months reveals a non-enhancing post ablative changes in the lower pole of the left kidney

In our case the patient initially presented with a mass in close relation to the Inferior Vena Cava. A very small percentage (2%) of LMS is of vascular origin. Seen more commonly in women in the sixth decade of life, these tumors usually manifest late in the course of the disease due to nonspecific symptoms\[8-10\]. IVC LMS usually end up being a post-surgical pathological diagnosis\[8\]. Despite high Recurrence, radical surgical en bloc resection is the mainstay of treatment for IVC LMS. Extensive vascular reconstruction techniques may be necessary. Adjuvant chemo radiotherapy has been shown to prolong disease-free survival rates\[8, 9, 11\]. IVC LMS have a relatively good prognosis as compared to other vascular LMS\[12\].

Our patient remained symptom and disease free for up to seven years from surgery for the IVC LMS. She presented with a shoulder mass after 7 years. Imaging at that time also revealed a mass in the left kidney. Percutaneous CT guided biopsy proved this to be an LMS.

Renal sarcomas represent a small number (1%-2%) of malignant kidney tumors. Of these 50% are LMS. In the literature, renal LMS are represented primarily as case reports or as components of larger series of renal sarcomas. These tumors generally arise from the renal capsule, smooth muscle tissue of the renal parenchyma, renal pelvis and intrarenal vessels. Clinical presentation is similar to the renal cell carcinoma i.e. pain, palpable mass, and hematuria. Imaging features too may be indistinguishable from renal cell carcinomas\[13, 14\]. As a result it is frequently diagnosed on histological examination. Even on histology a diagnosis of LMS should be made with caution. In occasions, it may be difficult to differentiate LMS from the sarcomatoid renal cell carcinoma. Although sarcomatoid carcinomas have a distinguishing malignant epithelial component, they also have the spindle-shaped atypical cells seen in LMS which makes differentiating them challenging. Moreover, some epithelial markers can be present in pure smooth muscle sarcomas, while some smooth muscle markers are positive in carcinomas. Since the prognosis for a renal sarcoma is particularly poor, differentiation from sarcomatoid renal cell carcinoma is necessary\[14-16\]. Renal LMS show high grade morphology and aggressive
biological behavior with poor prognosis \cite{13,15}. Management of renal LMS is not defined due to the limited literature on the subject \cite{17}. Surgical resection is the standard treatment for these lesions. Adjuvant therapies like chemotherapy and radiation have met with limited success. Even after chemotherapry, survival is poor \cite{15,18}. There is very limited literature on renal LMS to analyze prognosis. Deyrup et al studied 10 patients from 3 large institutions. They reported that the majority of renal LMS are intermediate or high grade with correspondingly poor prognosis even after radical surgery \cite{19}. In another larger series, Kendal et al used the population-based Surveillance, Epidemiology, and End Results (SEER) registry to study 95,935 cases of invasive cancer of the kidney and renal pelvis retrieved from the SEER registry to provide 112 cases of LMS. Overall survival in renal LMS was dependent on the stage of tumor and age at diagnosis. Analysis revealed that the overall survival curve for renal LMS was similar to transitional cell carcinoma, and better than clear cell carcinoma. Results from Kendal et al provided a more optimistic view of renal LMS than was previously thought \cite{20}.

Our case is unique because our patient presented with masses in the scapula, kidney and nodules in the lung after a significant disease free interval of seven years. The masses in the scapula and kidney were histologically proven to be LMS. Whether these lesions are blood borne metastasis from the IVC with a delayed manifestation or multicentric LMS is not entirely clear. LMS are known to hematogenously metastasize to the lungs, liver, bones and brain. Other sites of metastasis are relatively rare \cite{21-23}. Also, although other soft tissue sarcomas have been reported as metachronous, there no reports of metachronous LMS but there are few series of multicentric (synchronous). A genetic predisposition for dedifferentiation of the tissues or congenital and acquired immunodeficiency may promote the clustering of sarcomas in one person \cite{24-26}. In view of limited literature on this topic and our unique presentation, both the scapular region mass and renal mass could represent blood borne metastasis or could be a part of the metachronous tumor.

Management of the renal mass was also unique. Since the mass in the left kidney appeared to be growing in size on sequential follow up imaging, it was decided to treat the lesion. Due to age of patient and significant morbidities including a number of surgeries in the past, a multidisciplinary team involving surgical oncologist, urologist, medical oncologist and interventional radiologist decided to proceed with percutaneous RFA of the renal mass. Renal cell carcinoma, which is the most common malignant tumor affecting the kidneys is usually managed with surgery, which is considered as a gold standard. The minimally invasive percutaneous thermal ablative therapies like radiofrequency and cryoablative therapies are evolving as alternatives, especially in patients with older age and those who are considered to be at high risk for surgery.

In various reported series nearly 100% success has been reported in tumors around 3.5 with a single session of ablation. Larger tumors greater than 4 cm have met with some limited success and may have needed additional sessions of ablation. Complications are not uncommon after RF ablation of small renal masses, with an incidence ranging from 4% to 20.6%. Most complications are self-limited and minor, including hematuria and transient neuropathy related to thermal injury to the psoas or paraspinal muscle groups. However some serious complications can occur, including severe skin burns, ureteral strictures and urine leaks, and inadvertent thermal injury to adjacent structures. Most series concluded that RFA provides reasonable long-term oncological control. Some data suggested that RFA had outcomes similar to surgery \cite{27-35}. Use of RFA however to treat a LMS of the kidney has not been reported before. Our patient is doing well clinically with follow up imaging 7 months after treatment revealed complete ablation with no recurrence of tumor.

In summary, we present a case of LMS involving multiple organ sites and presenting at different time intervals. Whether these were metastasis of the primary LMS in the IVC or represent metachronous LMS is unclear. Although RFA has been used to treat renal cell carcinoma in the kidneys, our use of a percutaneous minimally invasive technique in this clinical setting is unique. Although 7 months of follow up is a short period, our case demonstrate that RFA application in similar tumours of the kidney offers the advantage of a good oncologic result (at least initially), provides an option in management of tumours in patients with comorbidities and possibly decreases volume of tumour favoring a future more oncologic excision. To summarize, RFA may be considered as a therapeutic options in management of LMS in solid organs, especially if it is small (less than 3 cm) in size and patient is a poor surgical candidate.
References


