A Rare Case Of Renal Sarcoma With The Review Of Literature

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Abstract

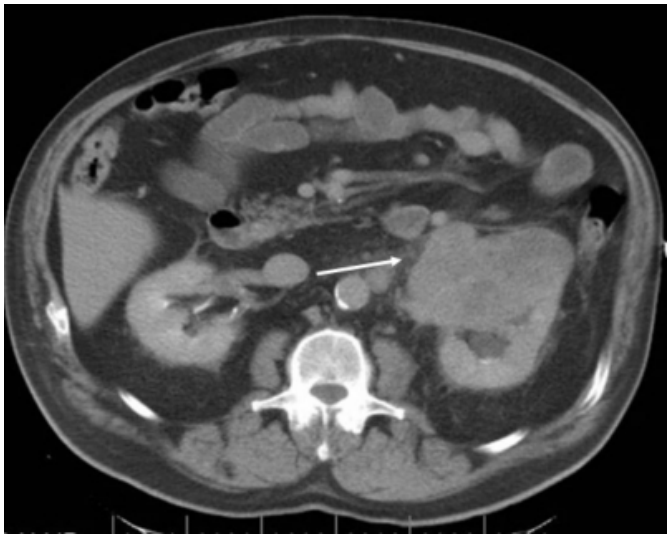
INTRODUCTION

Primary sarcomas constitute only 1%-2% of malignant renal tumors in the adulthood and are usually found as incidental tumors[1].Excluding Wilms tumors, leiomyosarcomas (LMSs) account for the majority of primary classifiable sarcomas arising in the kidney. Hence the clinical and treatment related information in the English literature is quite limited[2].

CASE REPORT

A seventy one year old male with morbid obesity had complaints of vague upper and left sided abdominal pain which was progressively getting worst for past six months. He also had experienced few episodes of hematuria in last two months in late 2009.He also had lost 20 pounds in the same time frame. At times he felt nauseous and quite weak. In February 2010, he had severe left sided abdominal pain not responding to regular non steroidal anti inflammatory drugs. The patient reported to local emergency. He had no other associated systemic symptoms. He had old history of myocardial infraction and obstructive sleep apnea. On clinical examination, the patient was very ill. There was an obvious swelling in left flank which seems to be moving with respiration. The mass was ballotable from the loin somewhat lobular to palpation. Urine analysis confirmed presence of red blood cells. Urine culture and cytology was negative for malignant cells. The basic blood work and renal panel was normal. Cardiac event was ruled out. Intravenous pyelogram showed normal right kidney but left kidney failed to concentrate the dye. The radiological appearances raised concern of a mass arising from the left kidney. CT scan of the abdomen confirmed a large heterogeneous mass arising from the left kidney measuring around 8.5 cm and metastatic foci within the left adrenal gland. There was evidence of lymphadenopathy medial to the kidney near the left renal hilum (Figure 1).

Figure 1



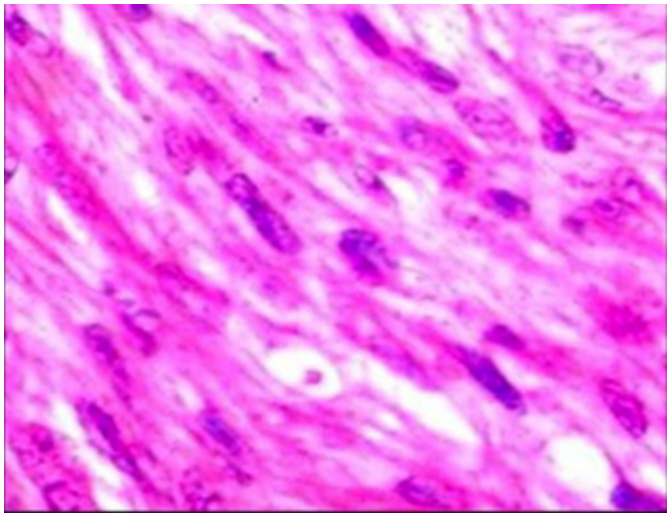
CT scan of chest and pelvis did not show any other metastases. CT scan of the brain and bone scan was normal. Differential at that point was a renal cell carcinoma at least Stage III because of the given fact that he had some lymphadenopathy. Subsequent to that the urologist recommended exploration with palliative left radical nephrectomy as the patient was continuously bleeding from the tumor mass as hemoglobin was dropping. A large left paramedian incision was made with transperitoneal approach. The highly vascular mass arising from left kidney was identified directly invading the left adrenal gland. The left nephrectomy was performed with difficulty as mass was adherent to posterior abdominal wall. The surgeon felt that there was a possibility of residual tumor left behind. In the postoperative period the patient did quite well.

The gross examination of specimen revealed an entire left kidney surrounded by large amount of fatty tissue. The entire specimen measured 24.0 cm from superior to inferior, 14.0

cm anteroposterior and 13.0 cm across and weighs 2267 grams. The specimen had a bosselated appearance. A segment of the ureter was identified measuring approximately 3.0 cm in length and 0.7 cm in its widest dimension. On section, the entire kidney was replaced by a bulging, pale gray, soft partly necrotic tumor. The tumor had a somewhat lobulated surface. Several tumor nodules were noted within the perinephric fat, measuring from 1.5 to 4.0 cm in dimension. The entire tumor within the kidney measured 13.0 x 6.0 x 6.5 cm. A small portion of the adrenal gland was identified and was involved by tumor. Tumor was extending into perinephric tissue, renal vein and segmental branches of vena cava above diaphragm. One regional node was identified which positive for malignancy was making it pT3bN1M0 by AJCC (American Joint Committee on Cancer) staging system.

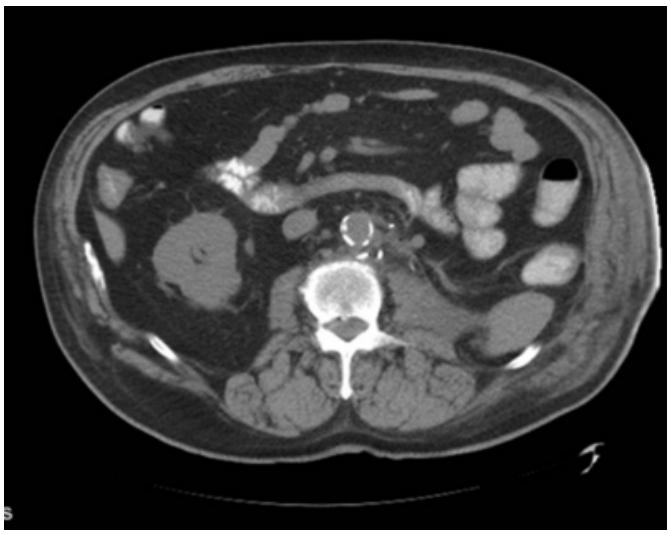
The tumor was composed of mainly spindle cells with areas of well-formed marked cellular pleomorphism. Typical renal cell carcinoma was not identified. The tumor was strongly positive for actin but negative for vimentin, CD10, pancytokeratin, CK7, HMB-45, Melanin A and desmin. The overall histological features and immunoprofile were compatible with leiomyosarcoma. Multiple tumor nodules were present within the perinephric adipose tissue but did not breach the Gerota's fascia. Left hilar tissue had the leiomyosarcoma present as well (Fig 2).

Figure 2



The patient was referred to tertiary cancer center to seek opinion about further adjuvant treatment. The case was discussed at length and literature was reviewed extensively at the local multidisciplinary tumor board meeting. Given the rare nature of disease and paucity of data, it was a challenge to formulate post operative adjuvant management. Since the patient had a positive margin with possibility of residual disease there was a speculation for high chances of local as well as distant relapse. In sarcomas such as in extremity with R1 or R2 resection, usually the post operative adjuvant radiation is strongly recommended in total dose of 66 to 70 Gy. However in retroperitoneal, there is limitation on total dose of radiation which can be safely administered because of normal tissue tolerance of small bowel and remaining kidney. After discussion and vigorous radiation treatment planning, patient received 54 Gray/30 fractions to left flank. The patient tolerated radiation quite well. The patient was offered systemic chemotherapy but he declined. At seven month post treatment follow up, the patient was clinically and radiologically doing very well (Fig 3).

Figure 3



DISCUSSION

Leiomyosarcoma of the kidney has preponderance in women and is more frequent in the fourth decade of life but can be found at almost any age, with a gradually increasing incidence in the later period of life[3]. Origin and histogenesis of leiomyosarcoma remains unknown till date. One of the strong hypothesis is renal sarcomas may arise from the smooth muscles fibers of renal parenchyma, renal capsule, renal pelvis or adjacent renal blood vessels[4]. Intravenous pyelography (IVP) most commonly reveals some kind of filling defect or even a calyceal deformity with or without hydronephrosis or poor functioning of the diseased kidney. Renal angiography findings of vascular pattern with increased vascularization and tortuous neoplastic vessels are non-specific and are quite similar to that of other intra-renal masses. Leiomyosarcoma can appear as a well-defined mass

of varying density and signal intensities on Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) respectively. The low signal intensity regions on T2 weighted MRI may exhibit delayed enhancement secondary to abundant fibrous tissue in the mass. These features may enable the experienced radiologist to refine the diagnosis of renal leiomyosarcoma considerably. However no imaging techniques can make absolute differentiation between leiomyosarcoma and renal cell carcinoma[5]. They usually present as solid or cystic masses. The most common symptoms and signs are abdominal pain, hematuria [6].

Grossly the tumors look like leiomyomas with a well-circumscribed margin and whorled cut surface. The malignant counterpart, however, appears fleshy and has areas of necrosis, hemorrhage and cystic degeneration. Leiomyosarcomas rarely metastasize to the kidney. In case of metastasis, they appear as intraparenchymal lesion or as a microscopic diagnosis. Bulky tumors that replace and invade the renal tissue are typical of renal leiomyosarcoma as seen in this case[7].

Microscopically, leiomyosarcoma show characteristics of smooth muscle tumor with alternating fascicles of spindle shaped cells. The cells have blunt ended, non tapering nuclei and eosinophilic cytoplasm. Indicators of malignancy are necrosis, nuclear pleomorphism and increase number of mitotic figures. Immunohistochemical examination usually show strong reactivity for actin and variable reactivity for vimentin, myosin, desmin, H-caldesmon, basal lamina components such as laminin and type IV collagen.[8]Retroperitoneal tumors with 5–10 mitosis per HPF are considered malignant. Tumors with 1–4 mitosis per HPF are potentially malignant if they are large and have areas of necrosis or nuclear atypia. Grignon et al. have recommended that large smooth muscle tumors should be treated with high suspicion unless proved otherwise. Focal myxoid change has also been reported[9].

Over all these tumors tend to have a poor prognosis. Majority of the patients develop distant metastasis despite use of systemic treatment. However in early tumors less than 5 cm, low histological grade, absence of lymph node metastasis and lymphovascular invasion with negative surgical margins may occasionally produce few long term survivors[10].

Radical nephrectomy has been suggested as the treatment of choice in the English literature[11]. Despite successful resection it usually shows an unfavorable prognosis .It eventually metastasizes to lungs, liver and colon. Most of the patients die within 24 months from diagnosis[12]. Because of the poor outcome of renal sarcomas, post operative adjuvant therapy with radiation and chemotherapy is an attractive choice for physicians. Brachytherapy in addition to external radiation has been tried but with little success. Unfortunately, no role for postoperative chemotherapy or radiotherapy has been established so far in the literature but few reports suggest that adjunctive radiation and chemotherapy may produce better outcome in terms of local control and possible benefit in over all survival[13].

CONCLUSION

In modern medicine with the help of sophisticated imaging modalities, leiomyosarcoma may be diagnosed pre-operatively. Strong suspicion about renal sarcoma should be raised in the lesion which exhibits a nodular mass with whorled cut surface. Histopathology and immunohistochemistry should be performed to establish correct diagnosis. Radical nephrectomy is the treatment of choice. Aggressive post operative adjuvant chemotherapy and radiotherapy may offer better results but not proven scientifically yet.

LEGENDS

Figure 1 Preoperative CT scan of the abdomen showing a large heterogeneous mass arising from the left kidney and within the left adrenal concerned for metastatic foci.

Figure 2 Microphotograph showing mainly spindle cells with areas of well-formed marked cellular pleomorphism. Haematoxlin and eosin stain 100x 100 magnifications.

Figure 3 Post operative CT scan of abdomen did not show any local recurrence.

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