INTRODUCTION

Renal leiomyosarcoma (LMS) is a rare form of aggressive renal cell cancer, which comprises approximately 1–3% of all adult malignant renal neoplasms arising from the renal capsule, renal pelvis or the vessel walls. They account for more than half of all the renal sarcomas. Most renal LMSs have conventionally been managed by radical nephrectomy with or without adjuvant radiotherapy and or chemotherapy. We report and describe one such case of capsular renal leiomyosarcoma encasing the inferior vena cava (IVC). We have reviewed and tabulated other such similar cases. The present case was successfully managed by radical nephrectomy and adjuvant radio-chemotherapy. Immunostaining should be freely used to define the histological type of renal sarcoma in order to accurately counsel and deliver a prognosis for patients with renal leiomyosarcomas with a poor prognosis.

CASE REPORT

A 50-year-old woman presented with a painful palpable right flank mass of six months duration. Ultrasonography revealed an irregular echogenic right kidney with a 60 × 74 mm homogenous mass lesion in the cortex of right kidney adherent to the liver and gallstones. Computerized tomography confirmed a large homogenous mass lesion involving the right kidney, encasing the infra-hepatic segment of the IVC closely abutting the liver [Figure 1a]. Metastatic work-up confirmed a localized renal mass. On exploration, the right renal mass was found to be adherent to and encasing a 5 cm length of infra-hepatic IVC [Figure 1b]. Right radical nephrectomy with excision and repair of the involved anterior wall of IVC was performed [Figure 1c]. There was no IVC tumor thrombus or hepatic involvement. Gross tumor [Figure 1d] and histopathology revealed right renal capsular LMS involving the renal capsule and pelvis, encircling the renal hilar vessels. The wall of the IVC was not grossly involved by the tumor and the ureter was also free. The H and E stained slide showed a uniform pattern of interlacing closely packed spindle cells on low power (10× - Figure 1e) with pleomorphic hyperchromatic elongated nuclei amidst an eosinophilic cytoplasm (HP-40× - Figure 1f), which suggested LMS. Immunohistochemistry revealed a negative (HMB-45 - Figure 1g) and cytokeratin (CK - Figure 1h) staining and an intensely positive cytoplasmic staining for smooth muscle actin (SMA - Figure 1i),...
confirming the diagnosis of LMS. The patient was referred for adjuvant chemotherapy and radiotherapy. At nine months of follow-up, the lady continued to be free from clinical signs of the disease.

**DISCUSSION**

Transitional cell carcinomas continue to be the commonest histological variety of renal pelvic neoplasms, followed by renal sarcomas. Renal sarcomas also remain a diagnostic rarity with renal LMS being most commonly reported and encountered subtype of renal sarcoma worldwide. Renal pelvic LMS are uncommonly reported with only four out of 30 cases of renal LMS being published and reported in the English literature to date [Table 1].

Renal LMS usually arise from renal capsule, renal pelvis or renal vessel smooth muscle fibers. It has been speculated though not proven that renal LMS arise from the renal capsule. They commonly present with flank pain and mass with or without hematuria. Occasionally they may present with spontaneous rupture and severe peri-renal hemorrhage. The diagnosis of LMS is generally suspected on H and E stained slides showing a monotonous population of spindle cells; however, this needs to be distinguished from the renal sarcoma and melanoma. Reliable differentiation from a sarcoma can be confirmed by immunohistochemistry (negative for antibodies to CK and HMB-45 and positive for antibodies to desmin and SMA; thereby indicating a mesenchymal neoplasm arising from muscle tissue), which also differentiates LMS from primary pure renal sarcomas, angiomyolipomas and nerve sheath cell tumors.

The reported comparative survival of renal LMS is about 25% (5-year overall survival) to 60% (5-year cause-specific survival) with a median overall survival of 25 months. Recurrences and metastasis are common. Renal LMS should
Table 1: The salient features of renal leiomyosarcomas reported to date

| Author            | No. | Presentation                                      | Management                  | FU  |
|-------------------|-----|---------------------------------------------------|------------------------------|-----|
| Demir et al. 2007 | 1   | Flank mass, pain, hematuria                       | NSS                         | 3 yrs|
| Sharma et al. 2007| 1   | Left flank pain, capsular LMS                     | RN, CT, SWRT                | 6½ yrs|
| Kartsanis et al. 2006 | 1 | Asymptomatic gross hematuria, pelvic LMS               | RNUT, no adjuvant therapy  | 3 yrs |
| Cocuzza et al. 2005 | 1 | Hypertension investigation                        | NSS (PN)                    | -    |
| Peyromaure et al. 2005 | 1 | Pelvic LMS                                        |                             | -    |
| Grasso et al. 2004 | 1  | Spontaneous rupture, flank pain, perirenal hemorrhage | RN                          | -    |
| Deyrup et al. 2004 | 10  | SMA, desmin, calponin (+)                          | Int-high grade, Mets (6) poor prognosis Recc (2) | -    |
| Moazzam et al. 2002 | 1  | Spontaneous rupture                                | RN+ splenectomy, high grade | -    |
| Moudouni et al. 2001 | 1 | Pelvic LMS                                        | Wide surgical excision      | -    |
| Dominici A et al. 2000 | 1 | Cystic LMS                                        | RN                          | -    |
| Kavantzas et al. 1999 | 3 | Flank pain, hematuria                              | RN                          | -    |
| El Otmany et al. 1999 | 1 | Flank pain                                        | RN+CT                       | Mets |
| Rebassa et al. 1999 | 1 | Pain, mass                                        | RN+RT                       | -    |
| Virseda et al. 1998 | 1  | Pain, mass                                        | RN+CT                       | -    |
| Lacquaniti et al. 1998 | 2 | Atypical clinical features                        | NSS                         | 6 yrs|
| Tamaki et al. 1994* | 1  | Pain, HDN, incidental tumor                        | RN                          | -    |
| Davis et al. 1992 | 1   | Pelvic LMS                                        | RN+RT+CT                    | 1 yr |

NSS - Nephron sparing surgery; RN - Radical nephrectomy; RNUT - Radical nephroureterectomy; HDN - hydronephrosis, PN - Partial nephrectomy; CT - Adjuvant chemotherapy; SWRT - Sandwich radiotherapy; FU - Followup

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Finnish renal leiomyosarcomas be treated as sarcoma using multimodality therapy so as to optimize survival and reduce tumor recurrence due to their overall poor prognosis. A review of similar cases of LMS [see Table 1] shows that in summary, renal LMS are aggressive tumors with poor prognosis. Radical nephrectomy has been the usual treatment of choice except for isolated cases presenting as localized small, polar renal nodules where a partial nephrectomy may suffice. A combination of aggressive radical surgical ablation with a selective anti-tumor signal blocking chemotherapy seems to be the preferred approach for managing most LMS.

The present report highlights the importance of deploying immunostaining in order to precisely define the exact histological type of renal sarcoma being encountered. This helps the treating urologist and surgeon in providing an accurate patient prognosis, with proper follow-up counseling and management.

REFERENCES

1. Demir A, Yazici CM, Eren F, Türkeri L. Case report: good prognosis in leiomyosarcoma of the kidney. Int Urol Nephrol 2007;39:7-10.
2. Chung YG, Kang SC, Yoon SM, Han JY, Seong DH. Leiomyosarcoma arising from the blind end of a bifid renal pelvis. Yonsei Med J 2007;48:557-60.
3. Chow LT, Chan SK, Chow WH. Fine needle aspiration cytodiagnosis of leiomyosarcoma of the renal pelvis. A case report with immuno-histochemical study. Acta Cytol 1994;38:759-63.
4. Kartsanis G, Dourous K, Zolota V, Perimenis P. Case report: leiomyosarcoma of the renal pelvis. Int Urol Nephrol 2006;38:211-3.
5. Ng WD, Chan KW, Chan YT. Primary leiomyosarcoma of the renal capsule. J Urol 1985;133:834-5.