INTRODUCTION

Leiomyosarcoma of the inferior vena cava (IVC) is a rare malignant tumor originating from the smooth muscle fibers of the media. Unlike intravenous leiomyomatosis which usually occurs in the veins of the myometrium and shows well differentiated smooth muscle cells with no or only a modest degree of nuclear pleomorphism and no or rare mitotic figures, leiomyosarcoma arises preferentially in the vena cava and usually shows high-grade tumors with easily recognized mitotic figures. Although only 218 cases of leiomyosarcoma of IVC have been reported in the international registry until 1994, leiomyosarcoma is the most common malignant tumor of the IVC (1). A complete resection of the tumor is the only treatment modality which may guarantee a long-term disease-free survival (2). When it comes to pararenal leiomyosarcoma of the IVC, ringed polytetrafluoroethylene (PTFE) graft interposition, and bilateral renal vein reconstructions in all patients. Postoperative radiation therapy was instituted in 3 of 4 patients. One patient who did not receive the postoperative radiation therapy was treated with adjuvant chemotherapy. The kidneys were preserved in all patients and no deep vein thrombosis (DVT) or venous insufficiency of the lower extremity veins developed. Distant metastasis to the lung was noted in one patient at 18 months after surgery, who was not received the postoperative radiation therapy but chemotherapy. In conclusion, a complete resection of the tumor, IVC reconstruction, and bilateral renal vein reconstruction followed by adjuvant radiation therapy is recommended for the treatment of pararenal leiomyosarcoma of the IVC.

MATERIALS AND METHODS

Between April 1999 and May 2002, the authors treated four patients with pararenal leiomyosarcoma of the IVC. We analyzed the clinicopathologic findings of these patients retrospectively.

Patients

All of the patients were referred to our hospital under the diagnosis of the retroperitoneal sarcoma on an abdominal computerized tomography (CT) scan (Fig. 1). Transfemoral caval biopsy was performed and the diagnosis of leiomyosarcoma of the IVC was made in one patient preoperatively. A laparotomy had been performed only for biopsy in one patient. All patients were female and the mean age was 50.8 yr (range 27 to 63 yr). All patients complained of upper abdominal pain for 12 to 36 months after surgery, who was not received the postoperative radiation therapy but chemotherapy. The kidneys were preserved in all patients and no evidence of distant metastasis in all patients.

Operation

In every case, we used an inverted T skin incision, mobilized the right and caudate lobes of the liver from the IVC, and prepared proximal and distal vascular control of the IVC at about 2-3 cm from the macroscopic tumor margins. Both the right and the left main renal veins were dissected. After
systemic heparinization, a proximal IVC clamping was performed step by step with monitoring the systemic blood pressure. Then, both renal veins and distal IVC were clamped. We routinely performed ringed polytetrafluoroethylene (PTFE) graft interposition and bilateral renal vein reconstructions; direct reimplantation of the right renal vein to the graft in all four patients, PTFE graft interposition between the left renal vein and the graft in three patients, and left renal vein ligation in one patient, following a complete resection of the tumor (Fig. 2). There was no systemic hypotension or circulatory collapse during the procedure in all patients. Some degree of renal congestion, especially in the right kidney, was noted intraoperatively but completely relieved on declamping in all cases. As a result, all kidneys were preserved and no postoperative renal functional impairment was noted (Fig. 3). Venovenous bypass and extracorporeal circulation techniques were not used in any of our cases.

Adjuvant therapy

All patients received postoperative adjuvant radiotherapy and/or chemotherapy. Radiotherapy at a does of 45-55 Gy was performed in three patients. Six cycles of chemotherapy (cyclophosphamide, doxorubicine, and dacarbazine) was given to two patients. One patient received both chemotherapy and radiotherapy.

RESULTS

Postoperative course

Postoperative course was uneventful in all patients except one, who required a second laparotomy for bleeding control on the first postoperative day. Kidneys were preserved in all patients and no deep vein thrombosis (DVT) or venous insufficiency of the lower extremity veins developed during the 6-41 months of follow-up period.

Pathology

All of the patients were considered to have a curative resection intraoperatively. One patient, however, showed a positive resection margin pathologically. The other patients had free tumor margins. The tumors ranged from 5 cm to 8.9 cm in greatest dimension (average, 7.05 cm) and formed nodular...
masses. All tumors showed a smooth muscle differentiation with interlacing fascicles and positive immunoreaction of smooth muscle actin and desmin. Three tumors showed a well differentiated histology and the remainder was a poorly differentiated tumor.

Recurrence

During 6 to 41 months follow-up period, a tumor recurrence was noted in one patient in the lung at 18 months after surgery, who was 27 yr old. The tumor showed a poorly differentiated type on pathology, and the patient had no adjuvant radiotherapy.

Table 1 shows the clinical findings and outcomes of all four patients.

**DISCUSSION**

Vascular leiomyosarcoma constitutes 2% of all leiomyosar-
4.0 8.9
2.5
3.8
4.5 8
6.8 5
4.3
2
60x729
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Clinical findings and outcomes of four patients with pararenal leiomyosarcoma of the IVC

| Patient | 1 | 2 | 3 | 4 |
|---------|---|---|---|---|
| Sex     | F | F | F | F |
| Age (yr) | 56 | 27 | 63 | 57 |
| Symptom Abdominal Pain | + | + | + | + |
| Diagnosis | CT scan | CT scan | CT scan | CT scan |
| Operation in 4 patients | Resection of the tumor, PTFE graft interposition (16 mm, ringed), Direct reimplantation of the right renal vein to the graft, and Left renal vein reconstruction ligation reconstruction reconstruction ligation reconstruction |
| Operative Complication | Bleeding that required a second laparotomy for control Bleeding that required a second laparotomy for control |
| Findings Mass size (cm) | 7.3 ± 0.8 x 3.5 | 8.9 ± 4.5 x 3.8 | 8 ± 6.8 x 4.3 | 5 ± 2.5 x 2 |
| Growth pattern Intraluminal | Dumbell | Dumbell | Dumbell |
| Margin involvement (-) | (-) | (-) | (+) |
| Cellularity ++ | ++ | +++ | ++ |
| Pleomorphism + | +++ | + | ++ |
| Necrosis (%) | 50 | 10 | 30 | 20 |
| Mitosis (No. of HPF*’s) | 10 | 7 | 20 | 5 |
| Adjuvant therapy Chemotherapy | yes | yes | no | no |
| Radiotherapy | yes | no | yes | no |
| Recurrence Local | no | no | no | no |
| Systemic | no | yes | no | no |
| F/U period (months) | 41 | 37 | 11 | 6 |
| Survival | yes | yes | yes | yes |

+, mild; ++, moderate; ++++, marked, HPF*: high power field.

Leiomyosarcoma of the IVC is reported to have a poor prognosis. Over half of patients (52.4%) who underwent a radical resection had recur at the median follow-up of 25 months (2, 9). Survival of patients with leiomyosarcoma of the IVC depends on the curative resection of tumor and its anatomical site. Involvement of the upper IVC segment and a high-grade tumor had a poor outcome whereas the middle segment tumor was associated with a better outcome (2, 9, 10). In our study, all four patients were considered to receive a curative resection intraoperatively but one patient showed a positive resection margin on pathologic examination. Histologic grades included three cases of well differentiated tumor and one case of poorly differentiated tumor. We performed adjuvant radiotherapy at a dose of 45-55 Gy to all patients except for one patient. Postoperative chemotherapy was given to two patients. Recurrence was noted in one patient in the lung at 18 months after surgery, who showed a poorly differentiated type of the tumor and received no radiotherapy. A controver-
sy exists in postoperative adjuvant therapy. Although we do not consider the postoperative adjuvant therapy routinely, a postoperative adjuvant radiation therapy seems to be beneficial for the control of the disease. In conclusion, leiomyosarcoma of the inferior vena cava is a rare malignant tumor, where a radical resection is imperative for securing a good prognosis. As for the pararenal leiomyosarcoma of the IVC, full mobilization of the right and caudate lobes of the liver is the key point to perform the curative resection of the tumor and preservation of the right kidney, and a gradual clamping of the proximal IVC under the monitoring the systemic blood pressure is the way to perform the procedure without venovenous bypass. Postoperative adjuvant radiation therapy seems beneficial to improve the survival of the patients with pararenal leiomyosarcoma of the IVC.

REFERENCES

1. Dzsinich C, Gloviczki P, van Heerden JA, Nagorney DM, Pairolero PC, Johnson CM, Hallett JW Jr, Bower TC, Cherry KJ Jr. Primary venous leiomyosarcoma: a rare but lethal disease. J Vasc Surg 1992; 15: 595-603.

2. Mingoli A, Feldhaus RJ, Cavallaro A, Stipa S. Leiomyosarcoma of the inferior vena cava: Analysis and search of world literature on 141 patients and report of three new cases. J Vasc Surg 1991; 14: 688-99.

3. Kevorkian J, Cento DP. Leiomyosarcoma of large arteries and veins. Surgery 1973; 73: 390-400.

4. Kulaylat MN, Karakousis CP, Doer RJ, Karamandjian HL, O’Brien J, Peer R. Leiomyosarcoma of the inferior vena cava: A clinicopathologic review and report of three cases. J Surg Oncol 1997; 65: 205-17.

5. Huguet C, Ferry M, Gavelli A. Resection of the suprarenal inferior vena cava: the role of prosthetic replacement. Arch Surg 1995; 130: 793-7.

6. Bower TC, Stanson AW. Tumors of the inferior vena cava: diagnosis and management. In Rutherford RB, ed: Vascular surgery, 5th ed, Philadelphia, 2000, WB Saunders.

7. Monig SP, Gawenda M, Erasmi H, Zieren J, Pichlmair H. Diagnosis, treatment and prognosis of leiomyosarcoma of the inferior vena cava. Three cases and summary of published reports. Eur J Surg 1995; 161: 231-5.

8. Yanaga K, Okadome K, Ito H, Matsumata T, Makino T, Okamura H, Sugimachi K. Graft replacement of pararenal inferior vena cava for leiomyosarcoma with the use of venovenous bypass. Surgery 1993; 113: 109-12.

9. Mingoli A, Cavallaro A, Sapienza P, Di Marzo L, Feldhaus RJ, Cavallani N. International registry of inferior vena cava leiomyosarcoma: analysis of a world series on 218 patients. Anticancer Res 1996; 16: 3201-5.

10. Burke AP, Virmani R. Sarcomas of the great vessels. A clinicopathologic study. Cancer 1993; 71: 1761-73.