EJVES Extra **14**, 19–21 (2007)
doi:10.1016/j.ejvsextra.2007.04.002, available online at http://www.sciencedirect.com on ScienceDirect

SHORT REPORT

Leiomyosarcoma of the Infrarenal Inferior Vena Cava: Management in Three Cases and a Review of the Literature

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Leiomyosarcoma (LMS) of the inferior vena cava (IVC) represents 90% of inferior vena cava tumours. It has a poor prognosis. We present three LMS of infrarenal IVC. Survival was long in two patients in spite of a non-radical resection. Chemotherapy and repeat surgery was performed for recurrence. The third patient was in complete remission at last follow-up at 29 months. Mainstay treatment for this tumour is surgical resection. Rescue surgery for local and metastastic recurrences together with systemic chemotherapy and radiotherapy may improve survival.

Keywords: Leiomyosarcoma; Inferior vena cava; Radical surgery; Rescue surgery.

Background

Only 0.5% of leiomyosarcomas (LMS) are of vascular origin and although rare, such lesions represent 90% of inferior vena cava (IVC) tumours. Fewer than 250 LMS of the IVC have been reported in the literature.[1,2] The clinical features are non-specific, leading to a delay in diagnosis. Radical surgery is the elective treatment for local control. However, the recurrences are frequent and the prognosis remains poor. We report three cases of IVC LMS.

Report

From 1995 to 2005, 56 patients with retroperitoneal sarcomas were treated at our hospital, a referral centre for these tumours. Three (5.3%) of these sarcomas were located in the infrarrenal IVC.

Clinical features and treatments are reported in Table 1. An infrarenal tumour of IVC was observed in all patients by echography, CT scan and MR angiography. Mean diameter was 9 cm. The preoperative histological study showed a high grade LMS (biopsy during a hysterectomy in patient 1 (P1) and FNAC in patient 2 (P2) and patient 3 (P3).

All patients were treated by surgery. P1 received chemotherapy (CT) prior to surgery. In all patients we performed marginal excision surgery. (Fig. 1) Partial resection of the IVC was performed in P1 and P2 and excision of the infrarenal IVC without reconstruction was performed in P3. Resection margins were positive in two cases and less than 1 cm in the other case. Postoperatively, all patients received radiotherapy (RT). Two patients (P1, P2) received postoperative CT (Doxorubicin and Ifosfamide).

Surgical complications were encountered in one patient: acute renal failure due to thrombosis of the right renal artery. This patient also presented oedema of the lower limbs due to the resection of vena cava; which resolved with compressive therapy.

Recurrences appeared at 24 months in two patients (P1, P2). All recurrences were treated with chemotherapy. P2 presented a solitary hepatic metastasis at 24 months that was treated with CHT followed by rescue surgery. At 12 months' follow-up, three hepatic metastasis were also treated with CHT and surgery. Sixty months after the initial surgery this patient presented a lung, bone and soft tissue dissemination and she died.

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Table 1. The three patients

	Sex	Age years	Clinical symptoms	CT-scan	Treatment	Margin	Adjuvant therapy	Recurrences	Treatment	Survival
P1	F	52	Dull hypogastric pain (1 yr duration)	Mass below celiac trunk	Surgery + suture v.cava	+	Preop CT Postop RT + CT	***24 mths:** Local + nodal mediastinic metastasis	CT	32 mths
P2	F	33	Mesogastric pain + alteration in intestinal motility (1 mth duration)	No invasion of neighbouring structures	Surgery + suture v.cava	<1 cm	Postop RT + CT	***24 mths:** hepatic metastasis ***36 mths:** hepatic progression metastasis ***60 mths:** lung, bone and soft tissue dissemination ***84 mths**: breast (metastasis) ***100 mths**: disease progression	*Repeated Surgery of recurrences *CT *RT	108 mths
P3	F	61	Right iliac fossa pain (2 yr duration)	No invasion of neighbouring structures	Surgery + resection of IVC and ligation of left renal vein	+	POSTOP RT	No recurrence	–	+29 mths free of recurrence

Fig. 1. Multilobulated tumour found in P3.

Death in the two patients was due to metastatic spread (Table 1). Although we were unable to perform radical surgery, P1 survived 32 months and P2 108 months. P3 is presently in complete remission 29 months since diagnosis.

Discussion

According to the literature,[3] surgery is the only treatment for LMS that significantly affects survival. Radical resection with disease-free margin of at least 1 cm is the recommended surgery but it is not always possible due to a delayed diagnosis. However, the developments in diagnostic techniques (CT-scan, MR) and their increased application enable earlier diagnosis and improve overall patient survival. RT and CHT can be used as adjuvant therapies although no survival increased has been demonstrated.

Hollenbeck *et al.*[3] compared survival in patients in two groups: patients who underwent radical resection and those who did not. Higher survival in the radical surgery group was statistically significant ($p < 0.05$). Hines *et al.*[4] presented 0% survival at 5 years in patients with positive margins. However, surgery of hepatic metastases has increased survival and it is possible to resect the mass and the metastases at the same time. Furthermore, debulking of intraabdominal recurrence offers good palliation.[5]

Although we could not perform radical surgery, survival was long in our three patients. One patient survived for 108 months. This patient underwent repetitive surgery of local recurrence and metastases

and the result was satisfactory. We consider that the relatively high survival in our patients was due to the aggressive surgical treatment of the recurrence.

Another controversial point is the reconstruction of the IVC. LMS usually grow slowly, so if there is involvement of the IVC with impairment of flow, collaterals will develop and when present, a resection can be performed safely. If the IVC is fully patent, it may be difficult to resect IVE without reconstruction. Before choosing a surgical technique, it is important to evaluate the previous patency of IVC, determine the segment to be resected, and assess the patient's general status.[6] We only encountered oedema in one patient and this was resolved with compressive therapy. Hollenbeck *et al.*[1,3] compared the various grades of oedemas using several techniques and concluded that simple ligature, primary reconstruction and autograft patch caused less oedema than prosthetic grafts. We consider that simple ligature or primary reconstruction is the first option with few complications and limited oedema due to marked collateral circulation.

In conclusion, with the increasing availability of non-invasive diagnostic techniques, LMS will be diagnosed earlier and more precisely, leading to a higher rate of radical surgery. Together with more aggressive treatment of recurrences and metastases, overall patient survival could improve. We consider that a multidisciplinary team composed of a medical oncologist, a radiotherapy oncologist, a general surgeon and a vascular surgeon can provide optimal treatment for this pathology.

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Accepted 27 April 2007