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CASE REPORT

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# Vascular leiomyosarcoma arising from the inferior vena cava diagnosed by intraluminal biopsy

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**Abstract** A 61-year-old woman developed pain in the right thigh, paraplagia of the lower extremities and lumbago in November 1996. A lumbar spine roentgenogram showed lytic change in L2, and magnetic resonance imaging showed a patchy destructive lesion and compression of the dural sac from the right by a tumour. Computed tomography (CT) myelography showed a motheaten destructive lesion in L2 and projection of the tumour into the spinal canal. Abdominal ultrasound, CT and cavography showed dilatation of the inferior vena cava (IVC) and an intraluminal tumour about 2×2.8×4 cm in size in the IVC. The tumour arose from the IVC just beneath the renal vein and extended to just short of the right atrium. Both vertebral and intraluminal biopsy materials showed the same morphology, in which atypical spindle cells admixed with multinucleated giant cells proliferated in a fascicular growth pattern. Neoplastic cells were strongly positive for alpha-smooth muscle actin. We diagnosed vascular leiomyosarcoma arising from the IVC with metastasis to the lumbar vertebrae. Cases of vascular leiomyosarcoma diagnosed by intraluminal biopsy are rare.

**Key words** Vascular leiomyosarcoma · Inferior vena cava · Intraluminal biopsy

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## Introduction

Leiomyosarcomas occur in the gastrointestinal tract, uterus, and soft tissues. In the soft tissues these tumours include cutaneous and subcutaneous types (superficial type), a retroperitoneal type, and a type of vascular origin. The most common presenting signs of vascular leiomyosarcoma are the Budd-Chiari syndrome, inferior vena cava (IVC) syndromes, and thrombosis [3, 4]. Mingoli et al. [13] recently reported an analysis of a worldwide series of 218 patients, including those known to the International Registry of inferior vena cava leiomyosarcomas. Generally, since these tumours often remain asymptomatic for a long period and may be difficult to diagnose, most cases are autopsy or operative diagnoses [12, 13]. Following the improvement of imaging techniques the detected incidence of vascular leiomyosarcomas has risen, but cases diagnosed by biopsy are very rare.

We report here a 61-year-old woman who suffered from pain, paraplagia and lumbago and was diagnosed by an intraluminal biopsy as having a vascular leiomyosarcoma arising from the IVC.

## Case report and pathological results

A 61-year-old woman underwent a simple hysterectomy because of leiomyoma of the uterine body when she was 56 years old. She developed pain in the right thigh, paraplagia of the lower extremities and lumbago in November 1996. She visited the orthopaedic department of Mito Saiseikai General Hospital on 28 May 1997

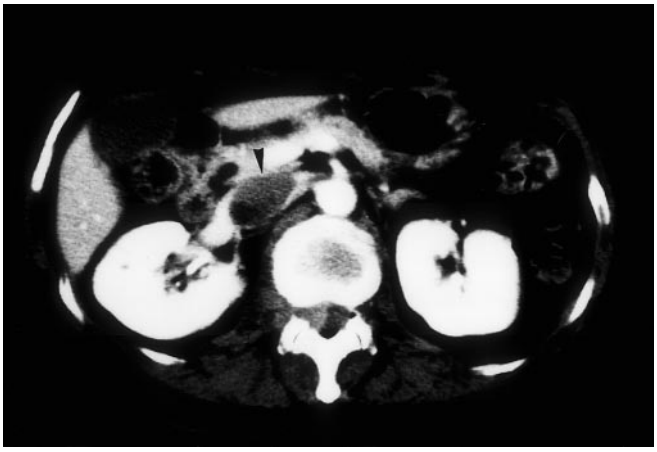

**Fig. 1** Computed tomography at the renal hilus or coeliac artery level. A luminal and bulky tumour (*arrowhead*) is observed in the dilated inferior vena cava. No tumour is observed in the retroperitoneum

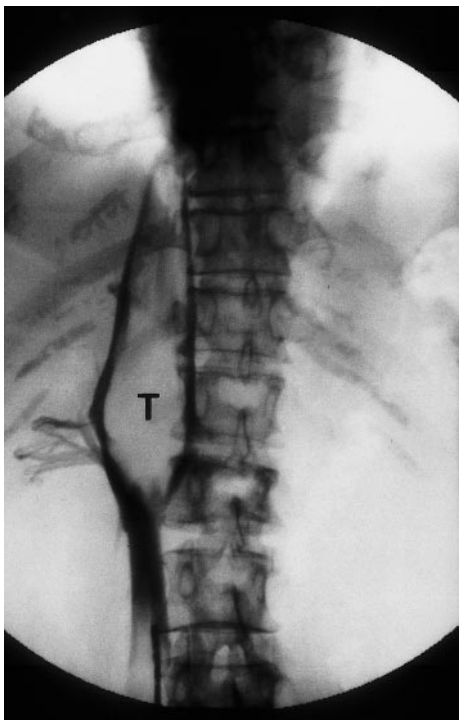


**Fig. 2** Cavography of the inferior vena cava (IVC) shows a luminal and slender cone-shaped tumour (*T*) that arose from the IVC just beneath the renal vein and grew to just before the right atrium

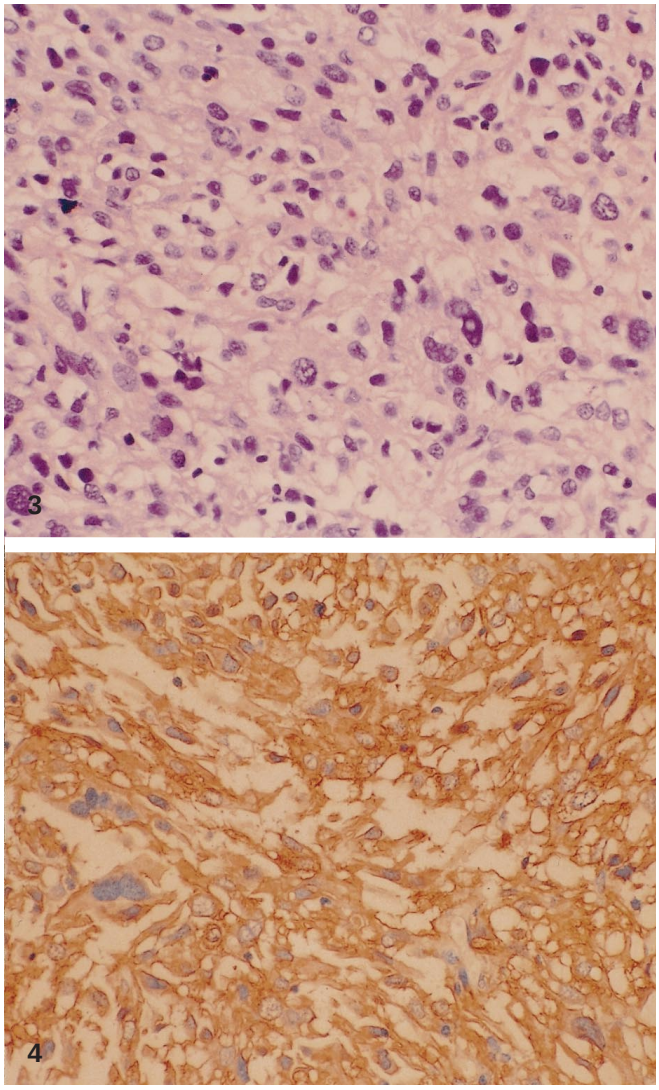


**Fig. 3** The second lumbar vertebral tumour sample taken by needle biopsy. Atypical spindle-shaped cells show a fascicular growth pattern admixing a few multinucleated giant cells. Haematoxylin & eosin, ×270

**Fig. 4** The luminal tumour in the inferior vena cava, as shown by an intraluminal biopsy. The neoplastic spindle cells were strongly positive for alpha-smooth muscle actin. Immunostain, ×270

because these symptoms had increased in intensity in April. She had marked tenderness on percussion over the spinous process of the second and third lumbar vertebrae (L2 and L3), hypaesthesia of the right L2 and L3 dermatomes, and absence of the right knee jerk. A lumbar spine roentgenogram showed lytic change of L2, which was confirmed by magnetic resonance imaging (MRI) on which compression of the dural sac by a tumour extending from the right side was evident. Computed tomographic myelography (CTM) also showed projection of the tumour into the spinal canal. A needle biopsy of L2 was performed on 6 June 1997, yielding a preliminary diagnosis of metastatic spindle-cell sarcoma (leiomyosarcoma or malignant schwannoma), because the tumour consisted of atypical spindle-shaped cells that had eosinophilic and fibrillary cytoplasm and showed a fascicular growth pattern admixed with a few multinucleated giant cells (Fig. 3). An investigation for the source of the sarcoma was made systematically. On 7 June 1997, the patient had urinary retention and progressive paraplagia and was given morphine when she developed severe pain. Abdominal ultrasound, computed tomography (CT) (Fig. 1), and cavography of the IVC (Fig. 2) showed dilatation of the IVC and a smooth surface, polypoid and intraluminal tumour (2×2.8×4 cm) arising in the middle segment of the IVC, extending from just beneath the renal veins to just short of the right atrium. The part of the tumour arising in the IVC at the renal vein level was bulky and the tail tapered gradually (Fig. 2). An intraluminal biopsy was made from the tumour of the IVC on 11 July 1997. This specimen (Fig. 4) had the same morphology as the lumbar vertebral tumour (Fig. 3). The neoplastic cells of both biopsy specimens were positive for alpha-smooth muscle actin (Fig. 4) and polyclonal actin, weakly

positive for vimentin, and negative for S-100 protein, myoglobin, and neuron-specific enolase. Based on the absence of a retroperitoneal tumour (Fig. 1), the presence of an intraluminal vascular tumour (Fig. 2), and morphological and immunohistochemical findings (Figs. 3, 4), we diagnosed the patient as having a vascular leiomyosarcoma arising from the IVC with metastasis to the lumbar vertebrae. An abdominal CT indicated two possible metastatic foci in the liver; however, a biopsy was not done. Bone scintigraphy showed a hot spot in L2 only. She was not treated with any therapy and died on 6 February 1998. Autopsy was not done.

## Discussion

This patient presented with orthopaedic symptoms. Several image analyses and immunohistochemical findings in an intraluminal biopsy revealed an intraluminal tumour considered to be a vascular leiomyosarcoma.

Burke and Virmani [4] reviewed 43 patients with sarcomas of the great vessels: 11 sarcomas of the aorta, 16 sarcomas of the IVC, and 6 sarcomas of the pulmonary artery. In their report, most of the leiomyosarcomas of the IVC occurred in women: the female-to-male ratio was 4.5 to 1. Sarcoma of the IVC was predominantly extraluminal in 14 of the 16 cases, and mostly intraluminal in only 2 cases. They reported a 76-year-old Caucasian male patient with a case similar to that of our patient, presenting with a metastasis to the femur for which a biopsy was performed. In their patient, the IVC tumour was found at autopsy. Of the 16 leiomyosarcomas of the IVC, 13 were resected with a portion of the IVC, 1 was removed as a thrombus only, and the remaining 2 were discovered at autopsy. In another report, of 62 cases, the diagnosis was made preoperatively in only 2 cases [1]. All cases of leiomyosarcoma of the IVC were autopsy or surgical cases [11], and the diagnosis had rarely been suspected before the operation [2]. The signs and symptoms are so nonspecific that 44% of the cases have not been diagnosed until at autopsy and 50% not until surgery [6]. Advances in imaging techniques have enabled a more accurate diagnosis prior to surgical resection [15] and have thus prolonged survival [9].

Parrilla et al. [14] reported a patient with leiomyosarcoma of the IVC whose diagnosis was confirmed before surgery by a needle biopsy. Coughlin and Andrews [8] reported a patient in whom a venous biopsy revealed a malignant spindle-cell neoplasm; surgical excision confirmed the presence of a leiomyosarcoma. Cases such as that of our patient, who was diagnosed as having leiomyosarcoma of the IVC by an intraluminal biopsy, are thus very rare. Similarly, Thompson et al. [16] reported a 54-year-old man suffering from bilateral leg oedema and a mass related to his IVC shown on CT. A transluminal biopsy obtained suggested the presence of a smooth muscle tumour. Cacoub et al. [5] recommended the diagnosis of leiomyosarcoma of the IVC by biopsy guided by ultrasonography or CT scan.

Retroperitoneal leiomyosarcomas are usually quite large and often unresectable. In Enzinger and Weiss's experience with 36 retroperitoneal leiomyosarcomas, the mean size was about 16 cm [10]. The retroperitoneal leiomyosarcoma commonly involves the kidneys, pancreas and vertebral body by direct extension [10]. In our case, CT and MRI performed on several occasions revealed no retroperitoneal mass between the IVC and vertebral column. The intraluminal vascular tumour was distinctly detected by CT (Fig. 1) and by cavography of the IVC (Fig. 2). We therefore considered that the present case was a vascular leiomyosarcoma arising from the IVC, and not a retroperitoneal leiomyosarcoma directly invading the IVC and vertebral column.

In general, the liver and lungs are most frequently involved in haematogeneous dissemination [7]. In our case, chest roentgenograms and CT were performed on several occasions without revealing any abnormal shadows in the lungs. Oncologists have experienced cases with carcinoma, especially of the gastrointestinal tract, that had metastasized to the adrenal glands, kidneys, bone, or subcutaneous tissue by haematogeneous spread with no evidence of metastasis to the liver and lungs. We considered that this intraluminal tumour arising had metastasized to the lumbar vertebral body via the bloodstream without implantation in the lungs. However, the possibility of microscopically detectable micro-metastases in the lungs cannot be excluded.

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