Primary Leiomyosarcoma in the Inferior Vena Cava Extended to the Right Atrium: A Case Report and Review of the Literature

Shuichi Fujita\textsuperscript{a} Hideaki Takahashi\textsuperscript{a} Yumiko Kanzaki\textsuperscript{a}
Tomohiro Fujisaka\textsuperscript{a} Yoshihiro Takeda\textsuperscript{a} Hideki Ozawa\textsuperscript{b}
Hiroko Kuwabara\textsuperscript{c} Takahiro Katsumata\textsuperscript{b} Nobukazu Ishizaka\textsuperscript{a}

\textsuperscript{a}Department of Cardiology, Osaka Medical College, Osaka, Japan;
\textsuperscript{b}Department of Thoracic and Cardiovascular Surgery, Osaka Medical College, Osaka, Japan;
\textsuperscript{c}Division of Pathology, Osaka Medical College, Osaka, Japan

Keywords
Leiomyosarcoma · Inferior vena cava · Right atrium

Abstract
A 38-year-old woman had developed an abdominal distention, lower extremity edema, and dyspnea. Imaging examination revealed a large mass in the right atrium which was connected to lesions within the inferior vena cava. Although complete resection of the mass was not possible, partial surgical tumor resection was performed to avoid pulmonary embolization and circulatory collapse. Leiomyosarcoma was diagnosed histologically, and chemotherapy (doxorubicin) followed by radiotherapy was started. By reviewing papers published in the past 10 years that included 322 patients, we also discuss the clinical presentations and prognosis of leiomyosarcoma in the inferior vena cava.
Introduction

Leiomyosarcoma is a rare, malignant mesenchymal tumor; only 218 cases had been reported as of 1996 [1]. Leiomyosarcoma of vascular origin often occurs in the inferior vena cava, and one that originated from the wall of the inferior vena cava was first reported by Perl [2] in 1871. In a previous study, it was found that patients with inferior vena cava involvement may present with lower extremity edema; however, symptoms may be nonspecific, and overall prognosis is reported to be poor, with a median survival of 2 years [3].

We herein report the case of a 38-year-old woman with primary leiomyosarcoma that was thought to originate from the inferior vena cava and to extend to the cardiac cavity. We also review case reports describing primary intimal sarcomas of the inferior vena cava from the past 10 years, and discuss the prevalence of symptoms and compare survival periods in this relatively recent literature.

Case Report

A 38-year-old female patient experienced abdominal distension, lower extremity edema, and dyspnea from January 20xx. Elevation of hepatic enzyme levels, ascites, and enlargement of the inferior vena cava were found, and therefore the patient was admitted to her former hospital. Further examinations revealed a mass in the right atrium which extended to the inferior vena cava and hepatic and renal veins; thus, she was referred to our department for further diagnosis and treatment.

On admission, her vital signs showed a blood pressure of 138/96 mm Hg and a heart rate of 92 beats/min. Her abdomen was round and edema was present in both of the lower extremities. Chest X-ray revealed a normal cardiothoracic ratio of 48%. Electrocardiography did not show any apparent abnormal findings (fig. 1); it showed a preserved left ventricular ejection fraction of 61% and a tumor 33.0 × 35.7 mm in size in the right atrium that continued to the cavity of the inferior vena cava.

Laboratory studies showed a white blood cell count of 11,550 cells/μl, a hemoglobin level of 14.3 g/dl, a platelet count of 32.0 × 10⁴ cells/μl, and a D-dimer level of 5.7 μg/ml. Alanine transaminase and aspartate transaminase levels were elevated to 464 and 509 U/l, respectively. The levels of carcinoembryonic antigen, α-fetoprotein, CA19-9, and CYFRA 21-1 were within normal limits, but those of CA125 (389.9 U/ml) and PIVKA-II (43 mAU/ml) were elevated.

Computed tomography and magnetic resonance imaging showed dilatation of the inferior vena cava and tumor occupation between the right atrium and the inferior vena cava (fig. 2a–d), ascites, and myoma of the uterus. Subsequent [18F]-FDG-PET ([18F]-fluorodeoxyglucose positron emission tomography) showed increased nuclear uptake only in the mass in the right atrium (fig. 2e, f). Coronary angiograms showed no apparent coronary artery stenosis, with small arteries feeding the tumorous lesion from the right coronary artery and the left circumflex artery (data not shown).

Although complete resection of the tumor was considered to be difficult, a surgical approach was taken for the purpose of amelioration of hepatic congestion and avoidance of circulatory shock. The heart was exposed through a median sternotomy, and a cardiopulmonary bypass was established with an ascending aortic arterial return and venous drainage through the cannulae, one placed in the superior vena cava and the other directly into the right atrium via the appendage. The systemic temperature was reduced to 24°C, after which
the circulation was interrupted and the venous blood drained into an oxygenator. The right atrium was opened down to the inferior vena caval orifice. The intra-atrial portion of the solid tumor was carefully dissected, so as to prevent embolization, and then excised. The tumor was fragmented and removed by applying hundreds of bites with pituitary rongeurs so that the caval junctions of all the hepatic veins could be fully recanalized. After full rewarming, the cardiopulmonary bypass was discontinued uneventfully. The postoperative course was uncomplicated.

The tumor in the right atrial cavity, which was 48 × 45 mm in size, showed necrosis, congestion, and edema (fig. 3a, b). Histologically, the tumor was composed of intersecting fascicles of abundant large spindle cells with markedly bizarre nuclei and numerous mitotic figures (10 per 10 high-power fields; fig. 3c). Necrosis and myxoid degeneration were also seen. Immunohistochemically, the tumor cells were positive for α-smooth muscle actin and desmin (fig. 3d, e), while they were negative for pancytokeratin, CD31, CD34, and myogenin. The Ki-67 labeling index was about 70% in the hot spot (fig. 3f). Together with the clinical appearance and immunological characteristics of the tumor, a diagnosis of primary leiomyosarcoma originating from the inferior vena cava was made.

The patient underwent radiotherapy (55 Gy/25 Fr), as well as chemotherapy comprising 3 courses of 60 mg/m² doxorubicin triweekly as first-line chemotherapy, which led to a partial response. Then, the patient was administered eribulin.

Discussion

We reported a case of leiomyosarcoma originating from the inferior vena cava and extending to the right atrium. Although complete resection of the tumor was difficult, surgical treatment was selected to avoid progression of hepatic failure, pulmonary embolization [4], and circulatory collapse. After the surgery, the patient was undergoing chemotherapeutic and radiotherapy and was followed up on an outpatient basis.

We also performed a PubMed literature search of the past 10 years by entering the search terms 'leiomyosarcoma' and 'inferior vena cava' [3, 5–120]. These terms returned 196 articles with 322 cases (table 1). As reported, women were affected about 2.5-fold more often than men, although the mean age did not differ between genders (data not shown). The most prevalent symptoms were pain and/or discomfort, most frequently in the abdomen, and only 5.8% of the patients did not have any symptoms related to the leiomyosarcoma. Thus, as has been done in our patient, ultrasonographic examination may facilitate the detection of leiomyosarcoma in the inferior vena cava in subjects with gastrointestinal symptoms or edema.

Survival depends on the tumor’s size, location, and complete surgical resection [39]; the efficacy of chemotherapy and radiotherapy is limited [109, 121]. In the current study, data regarding surgery was available for 233 patients. Those who underwent surgery (n = 217) had significantly better survival than those who did not (n = 16) (fig. 4a). When patients who were reported in the papers published between 2007 and 2012 (n = 115) were compared with those reported in the papers published between 2012 and 2016 (n = 127), the prognosis was significantly better by log-rank test (fig. 4b). The rate of surgical resection (either incomplete or complete) in the publication period between 2012 and 2016 (138/151; 91.4%) was found to be significantly higher than that in the period between 2007 and 2012 (137/162; 84.6%; p = 0.083 by χ² test). The median survival period for the publication period of 2007–2016 was 6.75 years. The 5-year survival rate has been reported to be approxi-
approximately 50% after complete en bloc resection [1, 122] in papers published in the last century; however, an improvement in prognosis in recent years is suggested by the literature review of the current study.

In a recent report, Lv et al. [117] summarized the reports on 30 vascular leiomyosarcoma cases with involvement of the heart that had been listed on PubMed for the past 20 years. Of the 30 cases, 14 had right ventricular involvement. The average age at onset was 53.6 years, and there was a female dominancy (67%). The mean follow-up survival time for patients with single cardiac cavity involvement was 15 months, which seems to be much lower than without cardiac cavity involvement.

In summary, we reported on a 38-year-old female patient suffering from abdominal distension, lower extremity edema, and dyspnea and diagnosed with leiomyosarcoma of the inferior vena cava extending to the right atrium. Multimodality imaging should be considered for patients with suspected symptoms – even if they are often nonspecific – for early diagnosis and therapy.

Statement of Ethics

The current case report was following the Guidelines of the Ethics Committee at Osaka Medical College.

Disclosure Statement

The authors declare that they have no conflicts of interest.

References

1. Mingoli A, Cavallaro A, Sapienza P, et al: International registry of inferior vena cava leiomyosarcoma: analysis of a world series on 218 patients. Anticancer Res 1996;16:3201–3205.
2. Perl L: Ein Fall von Sarkom der Vena cava inferior. Virchows Arch 1871;53:378–383.
3. Kapoor R, Bansal A, Sharma SC: Leiomyosarcoma of inferior vena cava: case series of four patients. J Cancer Res Ther 2015;11:650.
4. Gowda RM, Gowda MR, Mehta NJ, et al: Right atrial extension of primary venous leiomyosarcoma: pulmonary embolism and Budd-Chiari syndrome at presentation – a case report. Angiology 2004;55:213–216.
5. Kleisi T, Raissi SS, Nissen NN, et al: Cavo-atrial tumor resection under total circulatory arrest without a sternotomy. Ann Thorac Surg 2006;81:1887–1888.
6. Ameei S, Butany J, Collins MJ, et al: Leiomyosarcoma of the inferior vena cava. Cardiovasc Pathol 2006;15:171–173.
7. Bonura A, Saade C, Sharma P: Leiomyosarcoma of the inferior vena cava. Australas Radiol 2006;50:395–399.
8. Spinelli A, Schumacher G, Benckert C, et al: Surgical treatment of a leiomyosarcoma of the inferior vena cava involving the hepatic and renal veins confluences: technical aspects. Eur J Surg Oncol 2008;34:831–835.
9. Delis S, Triantopoulou C, Bakoyiannis A, et al: Leiomyosarcoma of the infrarenal portion of the inferior vena cava in a cirrhotic patient with hepatitis C. Abdom Imaging 2008;33:222–224.
10. Al-Rikabi A, Hussain AA, Buchler M, et al: Primary leiomyosarcoma of the inferior vena cava: report of a case diagnosed by fine needle aspiration cytology and confirmed by histopathologic examination. Acta Cytol 2007;51:477–479.
11. Guerrero MA, Cross CA, Lin PH, et al: Inferior vena cava reconstruction using fresh inferior vena cava allograft following caval resection for leiomyosarcoma: midterm results. J Vasc Surg 2007;46:140–143.
Ceyhan M, Danaci M, Elmalı M, Ozmen Z: Leiomyosarcoma of the inferior vena cava. Diagn Interv Radiol 2007;13:140–143.

Ito H, Horlick J, Bertagnolli MM, et al: Leiomyosarcoma of the inferior vena cava: survival after aggressive management. Ann Surg Oncol 2007;14:3534–3541.

Mayer F, Aebert H, Rudert M, et al: Primary malignant sarcomas of the heart and great vessels in adult patients – a single-center experience. Oncologist 2007;12:1134–1142.

Sulfat LP, Mazza L, Farina EC, et al: Leiomyosarcoma of the inferior vena cava. Report of two cases and review of the literature. Ann Ital Chir 2007;78:303–306.

Streukens SA, Scheltinga MR, Ebels J, et al: A patient with vague inguinal complaints due to a leiomyosarcoma of the inferior caval vein (in Dutch). Ned Tijdschr Geneeskd 2007;151:2574–2579.

Rodríguez Gómez L, Rodrigo-Rivera García J, Alvarez Costelo L, et al: Leiomyosarcoma of the inferior vena cava. Incident finding (in Spanish). Arch Esp Urol 2007;60:127–131.

Satheesan B, Subramaniam SR, Kathiresan N, Sunil BJ: Postoperative renal failure following inferior vena cava tumor resection with right nephrectomy: a case report and review of literature. Indian J Urol 2008;24:104–106.

Tranchart H, Carlioni A, Balzarotti R, et al: Leiomyosarcoma of the inferior vena cava involving the renal veins: a simple method of right renal vein reimplantation. J Vasc Surg 2008;47:209–212.

Reges R, Denardi F, Matheus W, et al: Primary leiomyosarcoma of the inferior vena cava: how should it be treated and the vein anatomy re-established? Int J Urol 2008;15:259–260.

Jenssen C, Siebert T, Bartho S: Leiomyosarcoma of the inferior vena cava. Diagnosis using endoscopic ultrasound-guided fine-needle aspiration biopsy (in German). Dtsch Med Wochenschr 2008;133:769–772.

Bertini R, Suardi N, Marone EM, et al: Pregnant woman presenting with a gross retroperitoneal mass: surgical treatment with caval replacement. Eur Urol 2008;54:677–680.

Tan GW, Chia KH: An unusual case of leiomyosarcoma of the inferior vena cava in a patient with a duplicated inferior vena cava. Ann Vasc Surg 2009;23:256.e13–e18.

Cho SW, Marsh JW, Geller DA, et al: Surgical management of leiomyosarcoma of the inferior vena cava. J Gastrointest Surg 2008;12:2141–2148.

Stauffer JA, Fakhre GP, Dougherty MK, et al: Pancreatic and multorgan resection with inferior vena cava reconstruction for retroperitoneal leiomyosarcoma. World J Surg Oncol 2009;7:3.

Singh A, Perwaiz A, Kakodkar R, et al: Leiomyosarcoma of the inferior vena cava (IVC). Indian J Surg 2009;71:48–49.

Storj J, Pasumaryth L: Leiomyosarcoma of the inferior vena cava – a rare cause of abdominal pain. Am J Med Sci 2009;337:369.

Narata M, Okahta Y, Abe K, et al: Primary leiomyosarcoma of the inferior vena cava: case report. Abdom Imaging 2010;35:481–484.

Alexander A, Reeders A, Raffel A, et al: Leiomyosarcoma of the inferior vena cava: radical surgery and vascular reconstruction. World J Surg Oncol 2009;7:56.

Caso J, Ségne J, Back M, et al: Circumferential resection of the inferior vena cava for primary and recurrent malignant tumors. J Urol 2009;182:887–893.

Tameo MN, Calligaro KD, Antin L, Dougherty MJ: Primary leiomyosarcoma of the inferior vena cava: reports of infrarenal and suprarenal caval involvement. J Vasc Surg 2010;51:221–224.

Zhang H, Kong Y, Zhang H, et al: Leiomyosarcoma of the inferior vena cava: case report and treatment of recurrence with repeat surgery. Ann Vasc Surg 2010;24:417.e5–e9.

Daylamir A, Amiri A, Goldsmith B, et al: Inferior vena cava leiomyosarcoma: is reconstruction necessary after resection? J Am Coll Surg 2010;210:185–190.

Chia-Hsin L: Education and imaging. Hepatobiliary and pancreatic Budd-Chiari syndrome secondary to leiomyosarcoma of the inferior vena cava. J Gastroenterol Hepatol 2010;25:218.

Hassan M, Giancino G, Shirodkar SP, et al: Surgical technique of removal of inferior vena cava leiomyosarcoma extending into the right atrium without deep hypothermic circulatory arrest. J Card Surg 2010;25:277–291.

Reddy VP, Vanveldhuizen PJ, Muehlebach GF, et al: Leiomyosarcoma of the inferior vena cava: a case report and review of the literature. Cases J 2010:3:71.

Sessa B, Iannicelli E, Caterino S, et al: Imaging of leiomyosarcoma of the inferior vena cava: comparison of 2 cases and review of the literature. Cancer Imaging 2010;10:80–84.

Matsuyama A, Hisaoa M, Hashimoto H: Vascular leiomyosarcoma: clinicopathology and immunohistochemistry with special reference to a unique smooth muscle phenotype. Pathol Int 2010;60:212–216.

Laskin WB, Fannin-Smith JC, Burke AP, et al: Leiomyosarcoma of the inferior vena cava: clinicopathologic study of 40 cases. Am J Pathol 2010;14:873–881.

Kyriaz MA, Stafyla VK, Batzokolalou I, et al: Surgical challenges in the treatment of leiomyosarcoma of the inferior vena cava: analysis of two cases and brief review of the literature. Ann Vasc Surg 2010;24:826.e13–e17.
Wachtel H, Jackson BM, Bartlett EK, et al: Resection of primary leiomyosarcoma of the inferior vena cava: a case report. Case Rep Oncol Med 2012;2012:631010.

Soejima Y, Matsumoto T, Shibake K, Maehara Y: Tube cavoplastic using autologous vein grafts for resected inferior vena cava reconstruction. Surg Today 2013;43:452–455.

Gómez García ME, Carbonell Castelló F, García Espinosa R, Viñals Larruga B: Massive obstruction of venous return due to a primary inferior vena cava tumour (in Spanish). Cir Esp 2013;91:e17.

Webb EM, Wang ZJ, Westphalen AC, et al: Can CT features differentiate between inferior vena cava leiomyosarcomas and primary retroperitoneal masses? AJR Am J Roentgenol 2013;200:205–209.

Meyer F, Weber M, Schuh HU, Hallouz Z: Mid-term, relatively tumor-stable outcome after an initially successful interdisciplinary surgical intervention with locally achieved R0 resection status including a multimodal therapeutic concept of a metastasized leiomyosarcoma of the inferior vena cava (in German). Wien Med Wochenschr 2013;163:295–302.

Sadri BA, Amine AM, Zeineb M, et al: Leiomyosarcoma of the inferior vena cava. Clin Pract 2013;3:e8.

Perisano C, Maffulli N, Colelli P, et al: Misdiagnosis of soft tissue sarcomas of the lower limb associated with deep venous thrombosis: report of two cases and review of the literature. BMC Musculoskeletal Disord 2013;14:64.

Ohman JW, Chandra V, Poultides G, Harris EJ: Iliocaval and aortoiliac reconstruction following en bloc retroperitoneal leiomyosarcoma resection. J Vasc Surg 2013;57:850.

Li Y, Wang Y, Liu B, et al: 125I brachytherapy seeds implantation for inoperable low-grade leiomyosarcoma of inferior vena cava. Korean J Radiol 2013;14:278–282.

Ueda J, Yoshida H, Mamada Y, et al: Surgical resection of a leiomyosarcoma of the inferior vena cava mimicking hepatic tumor. Case Rep Med 2013;2013:235698.

Zaenkert EK, Bruns CJ, Winter H, et al: Resection of sarcoma involving the intrahepatic vena cava: report of 2 cases from a specialized center. Ann Vasc Surg 2013;27:498.e9–e13.

Liu Y, Sun Y, Jiang Y, et al: A novel strategy of vascular reconstruction after radical resection of an inferior vena cava leiomyosarcoma. Ann Vasc Surg 2013;27:803.e1–e5.

Lovisetto F, Corradini C, De Gesare F, et al: Leiomyosarcoma of the inferior vena cava: incidentally detected. Ann Vasc Surg 2013;27:803.e15–e19.

Cina CS, Ricciói V, Passanisi G, et al: Computerized tomography and 3-D rendering help to select surgical strategy in leiomyosarcoma of the inferior vena cava. Updates Surg 2013;65:283–288.

Dallaz BZ, Smith B, Tefera G, Weber S: Surgical management of retroperitoneal leiomyosarcoma arising from the inferior vena cava. J Gastrointest Surg 2013;17:2166–2171.

Lee HM, Jeong DS, Park PW, et al: Surgical treatment for an invasive leiomyosarcoma of the inferior vena cava. Korean J Radiol 2013;14:278–282.

Araujo RL, Gaujoux S, D’Albuquerque LA, et al: End-to-end renal vein anastomosis to preserve renal venous drainage following inferior vena cava radical resection due to leiomyosarcoma. Ann Vasc Surg 2013;28:1048–1051.

Kumar S, Kumar A, Guleria S: Primary leiomyosarcoma of the juxtarenal inferior vena cava: a case report. Indian J Surg 2013;75:313–315.

Levi Sandri GB, Suljice L, Boudjema K, Meunier B: Hepatobiliary and pancreatic leiomyosarcoma of the inferior vena cava. J Gastroenterol Hepatol 2014;29:896.

Yo T, Taoka R, Hanasaki T, et al: Leiomyosarcoma of the inferior vena cava: a case report and review (in Japanese). Hinyokika Kiyo 2014;60:115–119.

Naphade PS, Raut AA, Hira P, et al: Leiomyosarcoma of the inferior vena cava. Arch Iran Med 2014;17:383–387.

De Luca GM, Gurraldo A, Marzullo A, et al: Fainting as an unusual presentation of a large inferior vena cava leiomyosarcoma. Phlebology 2015;30:492–495.

Moazeni-Bistgani M, Basravi M: Leiomyosarcoma of the inferior vena cava. Asian Cardiovasc Thorac Ann 2016;24:72–74.

Jones A, Aziz M: Renal vein reconstruction for primary leiomyosarcoma of the inferior vena cava. ANZ J Surg 2016;86:729–731.

Yamamoto T, Yagi S, Hashida H, et al: Long-term survival following resection of a leiomyosarcoma originating from the inferior vena cava (in Japanese). Nihon Shokakibyo Gakkai Zasshi 2014;111:1624–1631.

Wei N, Xu XD, Xu H, Zu MH: Inferior vena cava leiomyosarcoma confirmed by catheter suction biopsy during digital subtraction angiography. Int J Clin Exp Med 2014;7:2365–2368.

Wachtel H, Jackson BM, Bartlett EK, et al: Resection of primary leiomyosarcoma of the inferior vena cava [IVC] with reconstruction: a case series and review of the literature. J Surg Oncol 2015;111:328–333.
Lim JH, Sohn SH, SungYW, et al: Banked vena caval homograft replacement of the inferior vena cava for primary leiomyosarcoma [sic!]. KoreanJThoracCardiovascSurg 2014;47:473–477.

SinghN, ShvidasaniD, KarangutkarS: Rare case of primary inferior vena cava leiomyosarcoma on F-18 fluorodeoxyglucose positron emission tomography-computed tomography scan: differentiation from non-tumor thrombus in a background of procoagulant state. Indian JNuc Med 2014;29:246–248.

Chan G, Kroczak T, Drachenberg D: Leiomyosarcoma of the inferior vena cava with renal metastasis: an unusual case and diagnostic challenge. CanUrolAssoc J 2014;8:358–360.

Barison A, Pastormerlo LE, Mirtzzi G, et al: Leiomyosarcoma of the inferior vena cava in a patient with Budd-Chiari syndrome. Rev PortCardiol 2014;33:807–809.

Nascimento RL, Antón AG, Fernandes GL, et al: Leiomyosarcoma of the inferior vena cava: a case report. Radiol Bras 2014;47:384–386.

Monteagudo Cortecero J, Guirau Rubio MD, Payá Romá A: Leiomyosarcoma of the inferior vena cava: AIRP best cases in radiologic-pathologic correlation. Radiographics 2015;35:616–620.

Matić P, Vučurević G, Babić S, et al: Intra-cardiac extension of the inferior vena cava leiomyosarcoma with Budd-Chiari syndrome presentation: a case report. Srp Arh CelokLek 2015;143:71–73.

Takatsuki M, Eguchi S, Hashizume K, et al: Liver autotransplantation for an inferior vena cava tumor. Transplantation 2014;98:e92–e94.

Miles LF, Hu R, Jones RM, et al: Inferior vena cava resection and hemihepatectomy for leiomyosarcoma, utilizing cardiopulmonary bypass, in situ hepatic perfusion, and distal hypothermic circulatory arrest. J CardiothoracVascAnesth 2016;30:169–175.

Flores L, Ferrer J, Pages M, et al: Leiomyosarcoma of the inferior vena cava: feasibility of surgical resection. A report of two cases. RevEspEnfermDig 2015;107:458–460.

Sonoda H, Minamimura K, Endo Y, et al: Complete surgical resection of a leiomyosarcoma arising from the inferior vena cava. Case Rep Med 2015;2015:342148.

Ippolito D, Querques G, Drago SG, et al: Duodenocaval fistula in a patient with inferior vena cava leiomyosarcoma treated by surgical resection and caval polytetrafluoroethylene prosthesis. Case Rep Radiol 2015;2015:575961.

Illuminati G, Pizzardi G, Calio F, et al: Outcome of inferior vena cava and noncaval venous leiomyosarcomas. Surgery 2016;159:613–620.

Sulpice L, Rayar M, Levi Sandri GB, et al: Leiomyosarcoma of the inferior vena cava and noncaval venous leiomyosarcomas. Surgery 2016;153:161–165.

Imai K, Ito M, Kanetsuki K, et al: Resection of recurrent leiomyosarcoma of the inferior vena cava with extention into the right atrium and Z stent implantation: report of a case [in Japanese]. KyobuGeka 2015;68:1093–1095.

Kim SH, Lee SH, Kim HS, et al: Multidisciplinary treatment of inferior vena cava leiomyosarcoma. ANZJ Surg 2016;86:104–105.

Alkhalihi E, Greenbaum A, Langsfield M, et al: Leiomyosarcoma of the inferior vena cava: a case series and review of the literature. AnnVascSurg 2016;33:245–251.

Liu L, Li X, Zhang Y: Hepatobiliary and pancreatic leiomyosarcoma: unusual cause of inferior vena cava obstruction. JGastroenterol Hepatobat 2016;31:1384.

Lv Y, Pang X, Zhang Q, Jia D: Cardial leiomyosarcoma with multiple lesions involved: a case report. IntJClin ExpPathol 2015;8:15412–15416.

Moncayo KE, Vidal-Insua JJ, Troncoso A, García R: Inferior vena cava leiomyosarcoma: preoperative diagnosis and surgical management. SurgCaseRep 2015;1:35.

Yakupoglu A, Ulus S, Cantasdemir M: Leiomyosarcoma of the inferior vena cava confirmed by aspiration biopsy with a catheter during digital subtraction angiography. VascEndovascularSurg 2016;50:164–167.

Singh S, Siriwardana PN, Johnston EW, et al: Perivascular parenchymal extension of the ablation zone following liver microwave ablation. BMJ CaseRep Med 2016;2016:bcr2015212871.

Hollenbeck ST, Grobmyer SR, Kent KC, Brennan MF: Surgical treatment and outcomes of patients with primary inferior vena cava leiomyosarcoma. JAmCollSurg 2003;197:575–579.

Hines JO, Nelson S, Quinones-Baldrich WJ, Eller R: Leiomyosarcoma of the inferior vena cava: prognosis and comparison with leiomyosarcoma of other anatomic sites. Cancer 1999;85:1077–1083.
Fig. 1. Chest X-ray (a) and electrocardiogram (b) on admission.

Fig. 2. Clinical images. a, b Coronal (a) and transverse (b) sections of computed tomography (CT) images. A tumor is visible in the right atrium that is continuously present within the inferior vena cava (arrows). c, d Coronal (c) and transverse (d) sections of magnetic resonance images. e, f Coronal (e) and transverse (f) sections of PET/CT-merged images. Increased FDG uptake may be observed in the right atrium (arrows).
Fig. 3. Histological analysis of the tumor. a Macroscopic appearance of the tumor resected from the right atrial cavity. b Cut surface of the tumor. c Hematoxylin and eosin staining. d Staining for α-smooth muscle actin. e Staining for desmin. f Staining for Ki-67. Original magnification ×100.

Fig. 4. Kaplan-Meier curve of the survival of the patients reported on in the past 10 years. a Subcategorized according to whether surgery had been performed or not. Patients who had undergone surgery had a significantly improved prognosis when compared with their counterparts who had not been surgically treated. b Subcategorized according to the year of publication. p values were obtained with the log-rank test.
Table 1. Summary of the papers on leiomyosarcoma in the inferior vena cava published during the past 10 years

| Women/men/unknown, n | 229/92/1 |
|----------------------|----------|
| Mean age ± SD, years | 54.4±13.7 |
| Symptoms at presentation (n = 139), n (%) | |
| Pain or discomfort | 111 (79.9) |
| Edema | 27 (19.4) |
| Mass | 13 (9.4) |
| Weight loss | 12 (8.6) |
| Dyspnea | 9 (6.5) |
| Therapies, n (%) | |
| Chemotherapy and radiotherapy (n = 248) | |
| Chemotherapy alone | 60 (24.2) |
| Radiotherapy alone | 48 (19.4) |
| Both chemo- and radiotherapy | 21 (8.5) |
| Neither chemo- nor radiotherapy | 119 (48.0) |
| Surgery (n = 313) | |
| Surgical resection | 275 (88) |
| No surgery | 38 (12) |