

Case Report

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

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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.

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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 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/ μ l, a hemoglobin level of 14.3 g/dl, a platelet count of 32.0×10^4 cells/ μ l, and a D-dimer level of 5.7 μ g/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 ^{18}F -FDG-PET (^{18}F -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 re-warming, 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 chemo- 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 χ^2 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-

mately 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

- 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.
- Perl L: Ein Fall von Sarkom der Vena cava inferior. *Virchows Arch* 1871;53:378–383.
- Kapoor R, Bansal A, Sharma SC: Leiomyosarcoma of inferior vena cava: case series of four patients. *J Cancer Res Ther* 2015;11:650.
- 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.
- Kleisli 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.
- Ameeri S, Butany J, Collins MJ, et al: Leiomyosarcoma of the inferior vena cava. *Cardiovasc Pathol* 2006;15:171–173.
- Bonura A, Saade C, Sharma P: Leiomyosarcoma of the inferior vena cava. *Australas Radiol* 2006;50:395–399.
- 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.
- 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.
- 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.
- 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.

- 12 Ceyhan M, Danaci M, Elmali M, Ozmen Z: Leiomyosarcoma of the inferior vena cava. *Diagn Interv Radiol* 2007;13:140–143.
- 13 Ito H, Hornick JL, Bertagnolli MM, et al: Leiomyosarcoma of the inferior vena cava: survival after aggressive management. *Ann Surg Oncol* 2007;14:3534–3541.
- 14 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.
- 15 Suffat 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.
- 16 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 Geneesk* 2007;151:2574–2579.
- 17 Rodríguez Gómez I, Rodríguez-Rivera García J, Alvarez Costelo L, et al: Leiomyosarcoma of the inferior vena cava. Incidental finding (in Spanish). *Arch Esp Urol* 2007;60:1.127–1.131.
- 18 Satheesan B, Subramaniam SR, Kathiresan N, Sunil BJ: Postoperative renal failure following inferior vena caval tumor resection with right nephrectomy: a case report and review of literature. *Indian J Urol* 2008;24:104–106.
- 19 Tranchart H, Carloni 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.
- 20 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.
- 21 Jenssen C, Siebert C, 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.
- 22 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.
- 23 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.
- 24 Cho SW, Marsh JW, Geller DA, et al: Surgical management of leiomyosarcoma of the inferior vena cava. *J Gastrointest Surg* 2008;12:2141–2148.
- 25 Stauffer JA, Fakhre GP, Dougherty MK, et al: Pancreatic and multiorgan resection with inferior vena cava reconstruction for retroperitoneal leiomyosarcoma. *World J Surg Oncol* 2009;7:3.
- 26 Singh A, Perwaiz A, Kakodkar R, et al: Leiomyosarcoma of the inferior vena cava (IVC). *Indian J Surg* 2009;71:48–49.
- 27 Srour J, Pasumarthy L: Leiomyosarcoma of the inferior vena cava – a rare cause of abdominal pain. *Am J Med Sci* 2009;337:369.
- 28 Narata M, Okuhata Y, Abe K, et al: Primary leiomyosarcoma of the inferior vena cava: case report. *Abdom Imaging* 2010;35:481–484.
- 29 Alexander A, Rehders A, Raffel A, et al: Leiomyosarcoma of the inferior vena cava: radical surgery and vascular reconstruction. *World J Surg Oncol* 2009;7:56.
- 30 Caso J, Seigne J, Back M, et al: Circumferential resection of the inferior vena cava for primary and recurrent malignant tumors. *J Urol* 2009;182:887–893.
- 31 Tameo MN, Calligaro KD, Antin L, Dougherty MJ: Primary leiomyosarcoma of the inferior vena cava: reports of infrarenal and suprahepatic caval involvement. *J Vasc Surg* 2010;51:221–224.
- 32 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.
- 33 Daylami R, Amiri A, Goldsmith B, et al: Inferior vena cava leiomyosarcoma: is reconstruction necessary after resection? *J Am Coll Surg* 2010;210:185–190.
- 34 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.
- 35 Hassan M, Ciancio 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;25:277–281.
- 36 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.
- 37 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.
- 38 Matsuyama A, Hisaoka M, Hashimoto H: Vascular leiomyosarcoma: clinicopathology and immunohistochemistry with special reference to a unique smooth muscle phenotype. *Pathol Int* 2010;60:212–216.
- 39 Laskin WB, Fanburg-Smith JC, Burke AP, et al: Leiomyosarcoma of the inferior vena cava: clinicopathologic study of 40 cases. *Am J Surg Pathol* 2010;34:873–881.
- 40 Kyriazi MA, Stafyla VK, Chatzinikolaou 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.

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- 41 Liu C, Zheng Y, Yang X, et al: Surgical resection of the inferior vena cava for leiomyosarcoma. *Ann Vasc Surg* 2010;24:822.e11–e15.
- 42 Bieliūniene E, Kavaliauskiene G, Mitraite D, et al: Leiomyosarcoma of the inferior vena cava. *Medicina (Kaunas)* 2010;46:200–203.
- 43 Ulla M, Kohan A, Pekolj J, et al: Direct 64-row MDCT venography in the diagnosis of an inferior vena cava leiomyosarcoma. *Abdom Imaging* 2011;36:333–336.
- 44 Krüger T, Mustafi M, Nadalin S, Aebert H: Vena cava sarcoma with tumor embolus in the pulmonary artery: surgical treatment. *Ann Thorac Surg* 2010;90:642–644.
- 45 Stoian I, Piser IT, Kulcsar I, et al: Rare tumors of the heart – angiosarcoma, pericardial lipoma, leiomyosarcoma. Three case reports. *J Med Life* 2010;3:178–182.
- 46 Huang J, Liu Q, Lu JP, et al: Primary intraluminal leiomyosarcoma of the inferior vena cava: value of MRI with contrast-enhanced MR venography in diagnosis and treatment. *Abdom Imaging* 2011;36:337–341.
- 47 Tilkorn DJ, Hauser J, Ring A, et al: Leiomyosarcoma of intravascular origin – a rare tumor entity: clinical pathological study of twelve cases. *World J Surg Oncol* 2010;8:103.
- 48 Nayyar R, Panda S, Saini A, et al: Leiomyosarcoma of inferior vena cava involving bilateral renal veins: surgical challenges and reconstruction with upfront saphenous vein interposition graft for left renal vein outflow. *Indian J Urol* 2010;26:438–440.
- 49 Ganeshalingam S, Rajeswaran G, Jones RL, et al: Leiomyosarcomas of the inferior vena cava: diagnostic features on cross-sectional imaging. *Clin Radiol* 2011;66:50–56.
- 50 Mann GN, Mann LV, Levine EA, Shen P: Primary leiomyosarcoma of the inferior vena cava: a 2-institution analysis of outcomes. *Surgery* 2012;151:261–267.
- 51 Mu D, Wang D, Zhou K, Zhu B: Radiographic features of intraluminal leiomyosarcoma of the inferior vena cava: an atypical case report. *Abdom Imaging* 2011;36:586–589.
- 52 Munene G, Mack LA, Moore RD, Temple WJ: Neoadjuvant radiotherapy and reconstruction using autologous vein graft for the treatment of inferior vena cava leiomyosarcoma. *J Surg Oncol* 2011;103:175–178.
- 53 Al-Saif OH, Sengupta B, Amr S, Meshikhes AW: Leiomyosarcoma of the infra-renal inferior vena cava. *Am J Surg* 2011;201:e18–e20.
- 54 Angiletta D, Fullone M, Greco L, et al: Leiomyosarcoma of the inferior vena cava: resection and vascular reconstruction using a Dacron graft and an Adam DeWeese clip – three-year follow-up. *Ann Vasc Surg* 2011;25:557.e5–e9.
- 55 Di Benedetto F, D’Amico G, Montalti R, et al: Banked depopulated vena caval homograft: a new strategy to restore caval continuity. *Surg Innov* 2012;19:NP5–NP9.
- 56 Fiore M, Colombo C, Locati P, et al: Surgical technique, morbidity, and outcome of primary retroperitoneal sarcoma involving inferior vena cava. *Ann Surg Oncol* 2012;19:511–518.
- 57 Ramponi F, Kench JG, Simring DV, et al: Early diagnosis and resection of an asymptomatic leiomyosarcoma of the inferior vena cava prior to caval obstruction. *J Vasc Surg* 2012;55:525–528.
- 58 Jadhav SA, Atluri VS, Prajapati R, Satoskar RR: Leiomyosarcoma of inferior vena cava. *J Postgrad Med* 2011;57:332–334.
- 59 Hashimoto M, Kobayashi T, Tashiro H, et al: A huge metastatic liver tumor from leiomyosarcoma of the inferior vena cava: report of a case. *Surg Today* 2012;42:505–508.
- 60 Kawahara T, Nakagawa T: A case of leiomyosarcoma arising from the inferior vena cava. *Jpn J Clin Oncol* 2012;42:83.
- 61 Kim JT, Kwon T, Cho Y, et al: Multidisciplinary treatment and long-term outcomes in six patients with leiomyosarcoma of the inferior vena cava. *J Korean Surg Soc* 2012;82:101–109.
- 62 Lotze U, Reponova J, Muth G, et al: Leiomyosarcoma of the inferior vena cava extending into the right atrium. A rare differential diagnosis of a right atrial tumor with fatal outcome. *Herz* 2012;37:573–578.
- 63 Yadav R, Kataria K, Mathur SR, Seenu V: Leiomyosarcoma of inferior vena cava: a case series of four cases. *Indian J Pathol Microbiol* 2012;55:83–85.
- 64 Arkadopoulos N, Karmanioliou I, Ekonomopoulos N, et al: Combination of total abdominal inferior vena cava resection with a novel technique of left renal outflow restoration. *Surgery* 2012;152:142–143.
- 65 Saboo SS, Ramaiya N, Jacene H, et al: Synchronous small bowel and atypical primary leiomyosarcoma of inferior vena cava in a patient with *RB1* mutation. *Abdom Imaging* 2014;39:33–39.
- 66 Carvajal López F, García Domingo MI, Herrero Fonollosa E, et al: Inferior vena cava leiomyosarcoma with liver metastasis. Multi-organ resection with vascular reconstruction (in Spanish). *Cir Esp* 2013;91:394–395.
- 67 Fernandez HT, Kim PT, Anthony TL, et al: Inferior vena cava reconstruction for leiomyosarcoma of Zone I–III requiring complete hepatectomy and bilateral nephrectomy with autotransplantation. *J Surg Oncol* 2015;112:481–485.
- 68 Wang Q, Jiang J, Wang C, et al: Leiomyosarcoma of the inferior vena cava level II involvement: curative resection and reconstruction of renal veins. *World J Surg Oncol* 2012;10:120.
- 69 Chan AC, Chan SC, Yiu MK, et al: Technical considerations for radical resection of a primary leiomyosarcoma of the vena cava. *HPB (Oxford)* 2012;14:565–568.

- 70 Kato S, Tanaka T, Kitamura H, et al: Resection of the inferior vena cava for urological malignancies: single-center experience. *Int J Clin Oncol* 2013;18:905–909.
- 71 Drukker L, Alberton J, Reissman P: Leiomyosarcoma of the inferior vena cava: radical surgery without vascular reconstruction. *Vasc Endovascular Surg* 2012;46:688–690.
- 72 Prieto Muñoz I, Pardo Masferrer J: Scalp metastasis from leiomyosarcoma of the inferior vena cava sign as the first clinical sign: a case report. *Case Rep Oncol Med* 2012;2012:631010.
- 73 Soejima Y, Matsumoto T, Shirabe K, Maehara Y: Tube cavoplasty using autologous vein grafts for resected inferior vena cava reconstruction. *Surg Today* 2013;43:452–455.
- 74 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.
- 75 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.
- 76 Meyer F, Weber M, Schulz HU, Halloul 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.
- 77 Sadri BA, Amine AM, Zeineb M, et al: Leiomyosarcoma of the inferior vena cava. *Clin Pract* 2013;3:e8.
- 78 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 Musculoskelet Disord* 2013;14:64.
- 79 Ohman JW, Chandra V, Poultides G, Harris EJ: Iliacaval and aortoiliac reconstruction following en bloc retroperitoneal leiomyosarcoma resection. *J Vasc Surg* 2013;57:850.
- 80 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.
- 81 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.
- 82 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.
- 83 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.
- 84 Lovisetto F, Corradini C, De Cesare F, et al: Leiomyosarcoma of the inferior vena cava incidentally detected. *Ann Vasc Surg* 2013;27:803.e15–e19.
- 85 Cina CS, Riccioli 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.
- 86 Dull 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.
- 87 Lee HM, Jeong DS, Park PW, et al: Surgical treatment for an invasive leiomyosarcoma of the inferior vena cava. *Korean J Thorac Cardiovasc Surg* 2013;46:373–376.
- 88 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* 2014;28:1048–1051.
- 89 Kumar S, Kumar A, Guleria S: Primary leiomyosarcoma of the juxtarenal inferior vena cava: a case report. *Indian J Surg* 2013;75:313–315.
- 90 Levi Sandri GB, Sulpice L, Boudjema K, Meunier B: Hepatobiliary and pancreatic: leiomyosarcoma of the inferior vena cava. *J Gastroenterol Hepatol* 2014;29:896.
- 91 Yo T, Taoka R, Hanasaki T, et al: Leiomyosarcoma of the inferior vena cava: a case report and review (in Japanese). *Hinyokika Kyo* 2014;60:115–119.
- 92 Naphade PS, Raut AA, Hira P, et al: Leiomyosarcoma of the inferior vena cava. *Arch Iran Med* 2014;17:383–387.
- 93 De Luca GM, Gurrado A, Marzullo A, et al: Fainting as an unusual presentation of a large inferior vena cava leiomyosarcoma. *Phlebology* 2015;30:492–495.
- 94 Moazeni-Bistgani M, Basravi M: Leiomyosarcoma of the inferior vena cava. *Asian Cardiovasc Thorac Ann* 2016;24:72–74.
- 95 Jones A, Aziz M: Renal vein reconstruction for primary leiomyosarcoma of the inferior vena cava. *ANZ J Surg* 2016;86:729–731.
- 96 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.
- 97 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.
- 98 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.

- 99 Lim JH, Sohn SH, Sung YW, et al: Banked vena caval homograft replacement of the inferior vena cava for primary leiomyosarcoma [sic!]. *Korean J Thorac Cardiovasc Surg* 2014;47:473–477.
- 100 Singh N, Shivdasani D, Karangutkar S: Rare case of primary inferior vena cava leiomyosarcoma on F-18 fluorodeoxyglucose positron emission tomography-computed tomography scan: differentiation from nontumor thrombus in a background of procoagulant state. *Indian J Nucl Med* 2014;29:246–248.
- 101 Chan G, Krocak T, Drachenberg D: Leiomyosarcoma of the inferior vena cava with renal metastasis: an unusual case and diagnostic challenge. *Can Urol Assoc J* 2014;8:358–360.
- 102 Barison A, Pastormerlo LE, Mirizzi G, et al: Leiomyosarcoma of the inferior vena cava in a patient with Budd-Chiari syndrome. *Rev Port Cardiol* 2014;33:807–809.
- 103 Nascif RL, Antón AG, Fernandes GL, et al: Leiomyosarcoma of the inferior vena cava: a case report. *Radiol Bras* 2014;47:384–386.
- 104 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.
- 105 Matic P, Vučurević G, Babić S, et al: Intracardiac extension of the inferior vena cava leiomyosarcoma with Budd-Chiari syndrome presentation: a case report. *Srp Arh Celok Lek* 2015;143:71–73.
- 106 Takatsuki M, Eguchi S, Hashizume K, et al: Liver autotransplantation for an inferior vena cava tumor. *Transplantation* 2014;98:e92–e94.
- 107 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 Cardiothorac Vasc Anesth* 2016;30:169–175.
- 108 Flores L, Ferrer J, Pages M, et al: Leiomyosarcoma of the inferior vena cava: feasibility of surgical resection. A report of two cases. *Rev Esp Enferm Dig* 2015;107:458–460.
- 109 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.
- 110 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.
- 111 Illuminati G, Pizzardi G, Calio F, et al: Outcome of inferior vena cava and noncaval venous leiomyosarcomas. *Surgery* 2016;159:613–620.
- 112 Sulpice L, Rayar M, Levi Sandri GB, et al: Leiomyosarcoma of the inferior vena cava. *J Visc Surg* 2016;153:161–165.
- 113 Imai K, Ito M, Kanetsuki K, et al: Resection of recurrent leiomyosarcoma of the inferior vena cava with extension into the right atrium and Z stent implantation; report of a case (in Japanese). *Kyobu Geka* 2015;68:1093–1095.
- 114 Kim SH, Lee SH, Kim HS, et al: Multidisciplinary treatment of inferior vena cava leiomyosarcoma. *ANZ J Surg* 2016;86:104–105.
- 115 Alkhalili E, Greenbaum A, Langsfeld M, et al: Leiomyosarcoma of the inferior vena cava: a case series and review of the literature. *Ann Vasc Surg* 2016;33:245–251.
- 116 Liu L, Li X, Zhang Y: Hepatobiliary and pancreatic: leiomyosarcoma: unusual cause of inferior vena cava obstruction. *J Gastroenterol Hepatol* 2016;31:1384.
- 117 Lv Y, Pang X, Zhang Q, Jia D: Cardiac leiomyosarcoma with multiple lesions involved: a case report. *Int J Clin Exp Pathol* 2015;8:15412–15416.
- 118 Moncayo KE, Vidal-Insua JJ, Troncoso A, García R: Inferior vena cava leiomyosarcoma: preoperative diagnosis and surgical management. *Surg Case Rep* 2015;1:35.
- 119 Yakupoglu A, Ulus S, Cantasdemir M: Leiomyosarcoma of the inferior vena cava confirmed by aspiration biopsy with a catheter during digital subtraction angiography. *Vasc Endovascular Surg* 2016;50:164–167.
- 120 Singh S, Siriwardana PN, Johnston EW, et al: Perivascular parenchymal extension of the ablation zone following liver microwave ablation. *BMJ Case Rep* 2016;2016:bcr2015212871.
- 121 Hollenbeck ST, Grobmyer SR, Kent KC, Brennan MF: Surgical treatment and outcomes of patients with primary inferior vena cava leiomyosarcoma. *J Am Coll Surg* 2003;197:575–579.
- 122 Hines OJ, Nelson S, Quinones-Baldrich WJ, Eilber FR: Leiomyosarcoma of the inferior vena cava: prognosis and comparison with leiomyosarcoma of other anatomic sites. *Cancer* 1999;85:1077–1083.

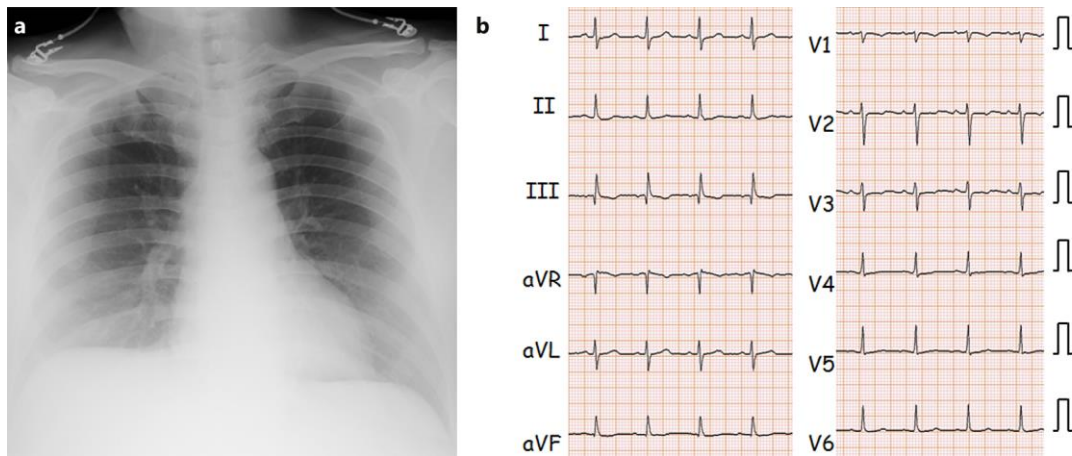


Fig. 1. Chest X-ray (a) and electrocardiogram (b) on admission.

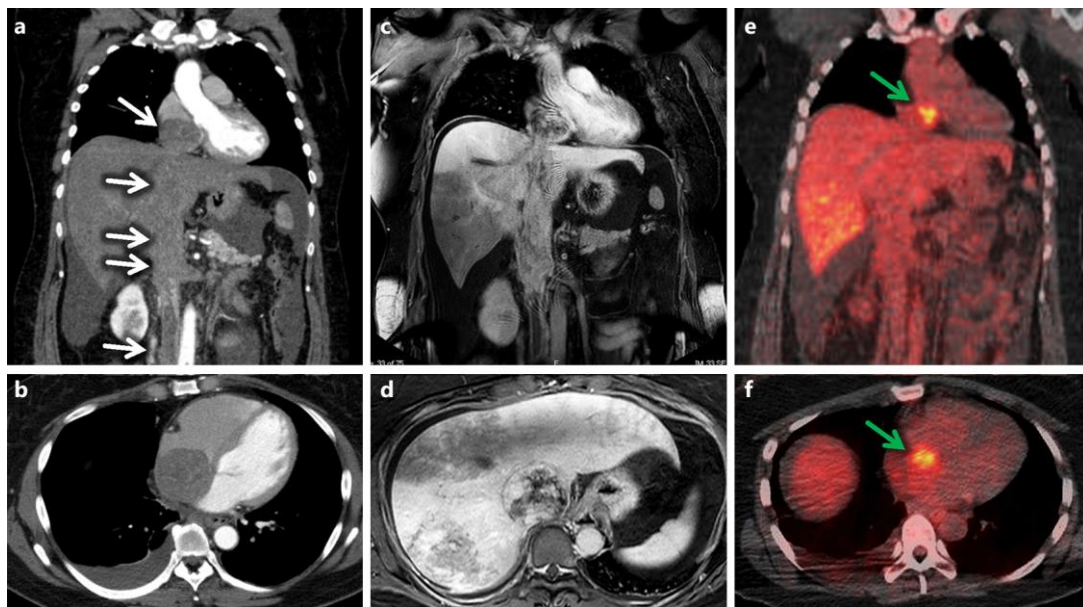


Fig. 2. Clinical images. **a, b** Coronal (a) and transverse (b) sections of computed tomography (CT) images. A tumor is visible in the right atrium that is continuously present within the inferior vena cava (arrows). **c, d** Coronal (c) and transverse (d) sections of magnetic resonance images. **e, f** Coronal (e) and transverse (f) sections of PET/CT-merged images. Increased FDG uptake may be observed in the right atrium (arrows).

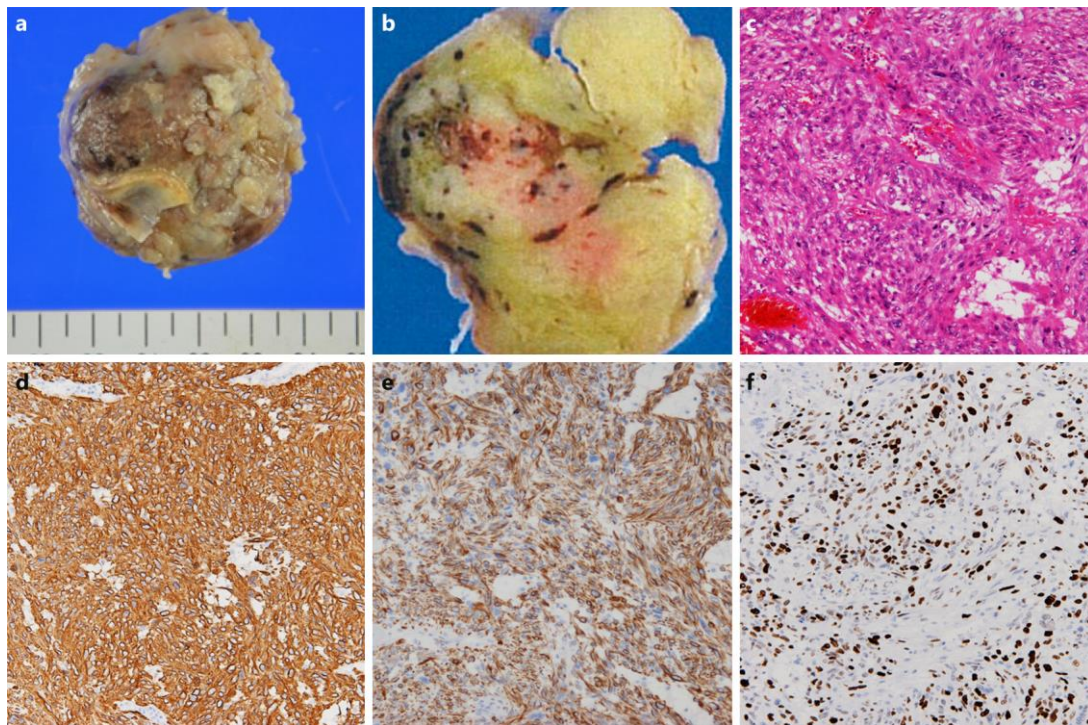


Fig. 3. Histological analysis of the tumor. **a** Macroscopic appearance of the tumor resected from the right atrial cavity. **b** Cut surface of the tumor. **c** Hematoxylin and eosin staining. **d** Staining for α -smooth muscle actin. **e** Staining for desmin. **f** Staining for Ki-67. Original magnification $\times 100$.

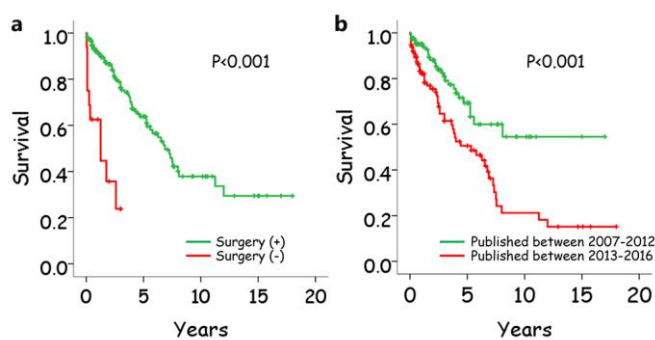


Fig. 4. Kaplan-Meier curve of the survival of the patients reported on in the past 10 years. **a** Subcategorized according to whether surgery had been performed or not. Patients who had undergone surgery had a significantly improved prognosis when compared with their counterparts who had not been surgically 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)