CASE REPORT

Preoperative diagnosis by three-dimensional angiography of a leiomyosarcoma arising from the left ovarian vein

Susumu Saigusa,^{1,2} Masaki Ohi,² Yasuhiro Inoue,² Masato Kusunoki²

¹Department of Surgery, Wakaba Hospital, Tsu, Mie, Japan
²Department of Gastrointestinal and Pediatric Surgery, Mie University Graduate School of Medicine, Tsu, Mie, Japan

Correspondence to

Dr Susumu Saigusa, saigusa@clin.medic.mie-u.ac.jp

SUMMARY

Leiomyosarcoma arising from the ovarian vein is extremely rare; we present a case with this unusual finding. A 78-year-old woman, diagnosed 2 years prior with a left retroperitoneal mass located in the lower part of the left kidney, was admitted to our hospital with a decrease in oral intake and a palpable, hard, abdominal mass. Contrast-enhanced CT showed a solid mass in the left lower abdominal cavity. On three-dimensional (3D) angiography the mass appeared to originate from the left ovarian vein. A simple total excision, including the involved vein, was performed and the tumour was found to be leiomyosarcoma. The patient's postoperative course was uneventful. There was no evidence of recurrence 5 months after surgery. In this patient, the previous information about the location of the retroperitoneal mass as well as the 3D angiography results were helpful in giving preoperative evidence of leiomyosarcoma originating from the left ovarian vein.

reported, both the previous information regarding the location of a retroperitoneal mass and three-dimensional (3D) angiography were helpful in providing preoperative decisive evidence of leiomyosarcoma arising from the left ovarian vein.

CASE PRESENTATION

A 78-year-old woman was admitted to our hospital with a decrease in oral intake and a left abdominal mass. At the age of 76 she had been diagnosed with a left retroperitoneal mass, measuring 60×46×50 mm, during an investigation for continuous low-grade fever (figure 1A). She declined surgical treatment at that time, despite the recommendation for total excision, as her fever resolved with administration of antibiotics and she had no abdominal symptoms. Physical examination on the recent admission revealed a large, hard, palpable mass, with poor mobility, in the left abdomen.

INVESTIGATIONS

On laboratory investigation, her inflammatory responses were elevated with a white blood cell count of 11 400/μl and C reactive protein level of 14.34 mg/dl (normal range <0.45 mg/dl); levels of tumour markers, including CA125, were within normal limits. Radiographic examination demonstrated intestinal gas shifted to the right side of the abdomen. Enhanced CT showed a mass in the left retroperitoneal space, with a diameter of 210×130×90 mm. This mass was circumscribed and heterogeneously enhanced (figure 1B). On 3D angiography, the left ovarian vein was seen to branch from the left renal vein and be adhered to the right dorsal portion of the mass. The inferior

BACKGROUND

Vascular leiomyosarcomas are rare lesions, representing less than 2% of all leiomyosarcomas; their prognosis is very poor compared with those that have other sites of origin.¹ The inferior vena cava is the most frequently observed site for leiomyosarcomas that arise from a vessel wall.² Excluding those of vena caval origin, vascular leiomyosarcomas most commonly originate in the renal vein, followed by the great saphenous vein, the pulmonary veins and the femoral vein³; leiomyosarcomas arising from the ovarian vein are extremely rare. The preoperative diagnosis of leiomyosarcoma and its origin is very difficult. In the patient, we

Figure 1 Axial contrast-enhanced CT images. (A) Retroperitoneal mass at initial diagnosis; the origin of the mass was unknown. (B) Left retroperitoneal mass is circumscribed and heterogeneously enhanced. There is no evidence of invasion into adjacent tissues.

To cite: Saigusa S, Ohi M, Inoue Y, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008823

Figure 2 Three-dimensional angiography showing the left ovarian vein branching from the left renal vein and adhering to the right dorsal portion of the mass. Left kidney and ovarian vein are shifted forward by the mass. Ao, aorta; IMA, inferior mesenteric artery; IVC, inferior vena cava; LOA, left ovarian artery; LOV, left ovarian vein; LRV, left renal vein; LU, left ureter; SMA, superior mesenteric artery; SMV, superior mesenteric vein; SV, splenic vein.

mesenteric artery was located in front of the mass and the left ureter was located dorsal to the mass (figure 2).

TREATMENT

A simple total excision, including the left ovarian vein, was performed. The operative findings confirmed that the mass occupied the left retroperitoneal space; it was covered by the peritoneum and there was a small amount of yellowish, serous ascites. The left ovarian vein was located immediately dorsal to the mass, flowing into it. There was no invasion of adjacent tissue; the left ureter was confirmed to be uninvolved. The mass, once removed, appeared ragged, greyish-white and solid, weighing approximately 2000 g (figure 3A).

Figure 3 (A) Resected mass after formalin fixation. Left ovarian vein is located dorsal to the mass. (B) Microscopic examination: leiomyosarcoma with 10 mitosis/10 high-powered fields (H&E stain; original magnification, $\times 200$).

OUTCOME AND FOLLOW-UP

On histopathological examination, the mass was determined to be leiomyosarcoma with 10 mitosis/10 high-powered fields (figure 3B). The patient's postoperative course was uneventful and she recovered well. Adjuvant chemotherapy was not planned given her advanced age and poor performance status. There was no evidence of recurrence at follow-up visit conducted 5 months after surgery.

DISCUSSION

Only eight cases of leiomyosarcoma arising from the ovarian vein have been reported so far in the literature.^{2 4–10} In the seven reports in which clinical findings were detailed,^{4–10} the mean age at initial diagnosis was 54 years (range 39–70 years), and the mean maximum tumour diameter was 7.3 cm (range 4–10 cm). Five masses (62.5%) originated from the right ovarian vein, with three originating from the left; three masses originated from the upper portion of the ovarian vein and three originated from the middle portion of the ovarian vein, with two originating from the lower vein. Our patient was the oldest at the time of initial retroperitoneal mass diagnosis, and the size of the resected lesion was larger, than in any previous reports. The chief complaints in leiomyosarcoma patients vary according to the location of the tumour. Although our patient had a known left retroperitoneal mass for 2 years, there were no abdominal symptoms such as colicky pain, despite the fact that the mass was gradually expanding. Hartman *et al*¹¹ reported that retroperitoneal leiomyosarcomas may be completely extravascular (62%), completely intravascular (5%) or with both extraluminal and intraluminal growth patterns (33%). Before pathological examination, our patient appeared to have the extravascular growth type.

The preoperative diagnosis of leiomyosarcoma arising from a blood vessel is very difficult to make. In this patient, if there had not been any previous information about the mass we would have made a preoperative diagnosis of a possible solid, malignant ovarian tumour. At the time of her initial presentation, the mass was near the lower portion of the left kidney but not attached to it and to the left of the left iliopsoas muscle and in front of the left ureter in the retroperitoneal space; the differential diagnosis was retroperitoneal paraganglioma, liposarcoma

or leiomyosarcoma. With this information we could rule out the possibility of an ovarian tumour; the findings suggested that the mass originated from the upper portion of the ovarian vein and 3D angiography confirmed that the left ovarian vein adhered to the right dorsal portion of the mass. Multidetector CT is a useful tool for the identification of the origin of vascular leiomyosarcomas.¹² Angiography demonstrates the degree of vascularity and the relationship between the mass and the surrounding blood vessels,¹¹ and multidetector CT is able to reconstruct an image using angiography data. 3D angiography by multidetector CT is of great benefit in the preoperative prediction of the tumour's origin.

Although Gage *et al*³ reported that 12% of vascular leiomyosarcomas, excluding those of vena caval origin, have metastases present at the time of initial diagnosis, our patient had no preoperative evidence of distant metastasis. We did not plan any adjuvant postoperative treatments as the patient was of advanced age and had a poor performance status. It has been reported that adjuvant treatment, such as anthracycline-based chemotherapy and combination chemotherapy with gemcitabine and docetaxel, is beneficial for survival; however, there is no evidence that adjuvant chemotherapy or radiotherapy improves the prognosis of patients with leiomyosarcoma.^{3 10} Although we observed no evidence of recurrence after surgery, careful follow-up of this patient is required because vascular leiomyosarcomas often do have distant metastases. These metastases are frequently seen in the liver, lung, brain and peritoneum; the metastatic risk is significantly higher in vascular leiomyosarcomas than in those that arise from other sites. In conclusion, the previous information about the location of the retroperitoneal mass as well as the 3D angiography results were helpful in giving preoperative evidence of leiomyosarcoma originating from the left ovarian vein in this case.

Learning points

- Leiomyosarcoma arising from the ovarian vein is extremely rare and its preoperative diagnosis is very difficult.
- The previous information about the location of the retroperitoneal mass as well as the three-dimensional (3D) angiography results were helpful in giving preoperative evidence of leiomyosarcoma originating from the left ovarian vein.
- 3D angiography is of great benefit in the preoperative prediction of a tumour's origin.

Contributors SS and MO conducted preoperative diagnosis and treatment of the patient. SS also contributed to writing the first draft of the manuscript. YI performed preoperative diagnosis and checking of the first draft of the manuscript. MK contributed to checking and correcting the manuscript draft. All the authors have read the manuscript and have approved this submission.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Caldarelli GF, Barellini L, Faviana P, *et al*. Leiomyosarcoma of the popliteal artery: case report and review of the literature. *J Vasc Surg* 2003;37:206–9.
- 2 Dzinich C, Gloviczki P, van Heerden JA, *et al*. Primary venous leiomyosarcoma: a rare but lethal disease. *J Vasc Surg* 1992;15:595–603.
- 3 Gage MJ, Patel AV, Koenig KL, *et al*. Non-vena cava venous leiomyosarcomas: a review of the literature. *Ann Surg Oncol* 2012;19:3368–74.
- 4 Honore LH, Robins RE, Taylor RH. Leiomyosarcoma of the right ovarian vein a case report. *Angiology* 1977;28:285–8.
- 5 Kawai K, Horiguchi H, Sekido N, *et al*. Leiomyosarcoma of the ovarian vein: an unusual cause of severe abdominal and flank pain. *Int J Urol* 1996;3:234–6.
- 6 Iannelli A, Karimjee BS, Fabiani P, *et al*. Leiomyosarcoma of the ovarian vein: report of a case. *Int Surg* 2003;88:6–8.
- 7 Cho HJ, Kim HK, Suh JH, *et al*. Leiomyosarcoma of the ovarian vein: a case report with radiological findings. *Korean J Radiol* 2008;9(Suppl):S14–17.
- 8 Chiarugi M, Pressi E, Mancini R, *et al*. Leiomyosarcoma of the right ovarian vein. *Am J Surg* 2009;197:e36–7.
- 9 Kim SH, Kwon HJ, Cho JH, *et al*. Atypical radiological features of a leiomyosarcoma that arose from the ovarian vein and mimicked a vascular tumour. *Br J Radiol* 2010;83:e95–7.
- 10 Tsuyoshi H, Yoshida Y, Kurokawa T, *et al*. Diagnosis and management of leiomyosarcoma arising from ovarian vein: case report and literature review. *J Obstet Gynaecol Res* 2012;38:466–70.
- 11 Hartman DS, Hayes WS, Choyke PL, *et al*. From the archives of the AFIP. Leiomyosarcoma of the retroperitoneum and inferior vena cava: radiologic-pathologic correlation. *Radiographics* 1992;12:1203–20.
- 12 Niver BE, Megibow AJ, Faust MJ, *et al*. Multidetector CT appearance of leiomyosarcoma of the splenic vein. *Clin Radiol* 2011;66:688–90.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit

<http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow