CASE REPORT

Successful removal of uterine leiomyosarcoma tumour thrombus propagating to the right atrium

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Accepted 7 November 2015

SUMMARY

Although intravenous leiomyomatosis is widely documented, intravenous extension of leiomyosarcoma into the inferior vena cava (IVC) and subsequently into the right atrium is extremely rare. Less than five such cases have been reported in the literature worldwide. Uterine leiomyosarcoma is an aggressive smooth muscle tumour occurring with an incidence of 1% in all female genital tract cancers and comprises about 3–7% of uterine cancers. It carries a generally poor prognosis with 5-year survival rates ranging from 18.8% to 65% across all stages. We report a case of primary uterine leiomyosarcoma with intravascular tumour propagation extending to the renal vein, IVC and right atrium of the heart, which was successfully resected in a one stage operation by a multidisciplinary team. This case demonstrates the importance of preoperative radiological staging and multidisciplinary planning.

BACKGROUND

This case clearly illustrates the need for multidisciplinary care in managing a patient with a complex and difficult problem.^{1–4} It is also an example of a rare disease, with only a handful of cases reported worldwide, hence it is a good experience to share for others to learn from.

CASE PRESENTATION

A 46-year-old woman, gravida 1/para 1, presented with abdominal distension associated with pelvic pain and abnormally heavy menstrual bleeding with clots. She also had symptoms of shortness of breath, giddiness and lethargy, which was attributed to anaemia; her haemoglobin level was 6.5 g/dL. Clinical examination revealed a relatively mobile 20 cm firm abdominopelvic mass. An endometrial biopsy confirmed a diagnosis of uterine leiomyosarcoma.

INVESTIGATIONS

Pelvic ultrasonography showed a 14 cm inhomogeneous hyperechoic uterine mass with high Doppler flow, with an associated 4.8 cm right ovarian cyst. MRI scan confirmed the pelvic mass to probably be a sarcoma, with possible tumour extension into the psoas muscle and enlarged left obturator lymph nodes (figure 1). The CT scan revealed a pelvic mass with tumour extension into the left ovarian vein, left renal vein, inferior vena cava (IVC) and the right atrium of the heart. There were no demonstrable tumour pulmonary emboli or distal metastases (figures 2 and 3).

Figure 1 Transverse section of MRI of the pelvis showing a large mass arising from the uterus, with involvement of the psoas muscle.

Transthoracic echocardiography showed a tumour mass in the right atrium prolapsing through the tricuspid valve annulus (figures 4 and 5).

TREATMENT

Although leiomyosarcoma generally carries a poor prognosis, the decision to proceed with urgent surgery was based on the high risk of sudden death from tumour emboli. Our patient was young, with good performance score, and the disease was highly likely to be completely resected. A multidisciplinary team, comprising of a gynaecologist, urologist and cardiothoracic surgeon, reviewed the histology and radiological staging, and planned the

Figure 2 Coronal section of CT of the abdomen showing the pelvic mass extending into the left ovarian vein, left renal vein and inferior vena cava (IVC).

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To cite: Lim YH, Lee LS, Lim CH, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-213384

Figure 3 Coronal sections of CT of the thorax showing the tumour thrombus in the right atrium of the heart.

operation with cardiac anaesthetic support. It was planned as a one stage operation.

A midline laparotomy and sternotomy approach was used to ensure good surgical exposure. The patient underwent intraoperative real time transoesophageal echocardiography. There was a 20-week-sized uterus with obvious tumour extending and

invading into the left gonadal vein, which was dilated to 3 cm diameter. The tumour thrombus extended and invaded into the left renal vein, IVC and supradiaphragmatic IVC. At the level of the renal vein, there was extravascular extension and invasion into the renal pelvis, psoas muscle and para-aortic nodes. The left distal ureter was encompassed into the tumour mass (figure 6).

We proceeded with total abdominal hysterectomy bilateral salpingo-oophorectomy including resection of the distal left ureter, which was completely encased in the tumour (figure 7). The left gonadal vessels were mobilised and ligated only on completion of hysterectomy, to reduce risk of tumour propagation. The enlarged left common iliac lymph nodes were resected. The IVC was skeletonised after mobilising the right lobe of liver by dividing along the triangular ligament and ligating the caudate veins (figure 8). The renal veins were skeletonised (figure 9). After cannulating the ascending aorta and superior vena cava, the patient was put on cardiopulmonary bypass. The left pulmonary artery was cannulated for full bypass and the patient was cooled down to 20°C to begin hypothermic circulatory arrest. Cavotomy up to the level of the right atrium showed that the tumour ascending from the gonadal vein was adherent to the renal vein and inferior vena cava. The caval component was peeled off the endothelium but the renal vein had to be sacrificed. The tumour was removed en bloc along with the renal and gonad vein, measuring a length of 45 cm (figure 10). A mobile piece of the tumour that had broken off was removed from the right atrium. The atrium and IVC were

Figure 4 Transthoracic echocardiography showing the tumour thrombus in the right atrium. IVC, inferior vena cava; LA, left atrium; SUV, superior vena cava; RA, right atrium.

Figure 5 Transoesophageal echocardiography showing tumour mass in the right atrium prolapsing through the tricuspid valve annulus.

Figure 6 Tumour replacing and distending the entire left gonadal vein and encompassing the left ureter.

reconstructed, and the patient was weaned off bypass uneventfully and warmed to normal body temperature. Total duration of cardiopulmonary bypass was 117 min and total circulatory arrest time was 14 min.

The patient was monitored in the cardiothoracic surgical intensive care unit overnight, and made an uneventful recovery and was discharged on postoperative day 10.

OUTCOME AND FOLLOW-UP

Final histology revealed stage 4B primary uterine leiomyosarcoma with lymphovascular invasion, metastatic tumours in the gonadal vein, IVC and right atrium, and involvement of the para-aortic lymph nodes.

Following her recovery, we gave the patient adjuvant chemotherapy including six cycles of gemcitabine and paclitaxel. Her surveillance CT scan showed no residual and no recurrent cancer. We are, at present, planning adjuvant radiotherapy at the para-aortic level where there is extravascular extension of the cancer. The patient is currently on her sixth dose of chemotherapy.

DISCUSSION

Intravenous leiomyosarcomatosis is an extremely uncommon condition; it can cause tumour pulmonary embolism that is often fatal.⁵ The most common tumour type associated with tumour invasion into the IVC is renal cell carcinoma, with 4–10% of patients having extension of tumour into the IVC via the renal vein. Other reported tumour types associated with venous invasion include retroperitoneal sarcomas, hepatocellular

Figure 7 Posterior view of hysterectomy specimen showing the primary leiomyosarcoma tumour.

Figure 8 View of the tumour filling the inferior vena cava (IVC) after mobilising the IVC off the liver.

carcinomas and adrenal cortical carcinomas.^{6,7} There are two theories with regard to the pathogenesis and development of intravenous leiomyosarcomatosis, the malignant subtype of intravenous leiomyomatosis. One proposed theory is that the intima of myometrial sinuses is invaded by leiomyomatosis cells of uterine myometrium. The alternative theory is that the malignancy is derived from proliferating smooth muscle cells originating from the venous walls of the uterine or pelvic veins.⁸

Preoperative diagnosis is very rare as it is usually diagnosed on hysterectomy specimen. Moorjani *et al*,⁹ however, reported a novel way of obtaining tissue diagnosis by performing a biopsy of the mass in the right atrium via the superior vena cava with transoesophageal guidance. High-resolution CT and MRI scans are useful to determine the nature and size of the tumour mass, and to evaluate the extent of tumour extension into the venous system with impact on staging, therapy and prognosis. Surgical planning can also be more directed after review of the scan images by an experienced radiologist. In our case, the patient was counselled about the need for cardiopulmonary bypass and circulatory arrest in order to achieve optimal surgical resection, which involved removing the tumour in the right atrium and IVC. Successful therapy can only be achieved with well-planned multidisciplinary care and effective communication with the ancillary staff.^{9,10} The operation theatre was reserved for the entire day and there were adequate blood products such as packed cells, platelets and fresh frozen plasma available in the event of massive blood loss. Experienced nurses and perfusionists were also required to assist in the surgery and management of the patient perioperatively.¹⁰

Figure 9 View of the liver, gall bladder and tumour filling the left gonadal vein, extending upwards to involve the left renal vein and IVC.

Figure 10 The distended gonadal vein filled with tumour on the left extending to a length of 20 cm, and the tumour that was removed en bloc from the inferior vena cava (IVC) and right atrium, measuring 25 cm in length.

No treatment guidelines exist for leiomyosarcomas as they are a rare group of gynaecological cancers,¹¹ and the occurrence of intravenous leiomyosarcomatosis that we have described has only been reported in a small number of case reports in the current literature. Learning points can be adapted from renal cell carcinoma with intracaval extension, as it behaves in a similar fashion. Concurrent and complete surgical excision of the primary as well as caval disease remains the standard of care for these patients.^{6 7 12} The role of adjuvant chemotherapy, radiotherapy and hormonal therapy remains unclear in patients with leiomyosarcoma, as long-term outcomes and data are lacking.^{1 13}

Although the prognosis for uterine leiomyosarcoma is poor, we felt that this extensive surgery was necessary to avert sudden death for this young, otherwise well woman. Tumour resection

Learning points

- Intravenous leiomyosarcomatosis with tumour thrombus propagating to the right atrium of the heart is a rare condition with less than five reported cases worldwide.
- Preoperative imaging is of utmost importance to help with surgical planning.
- Execution of complex surgeries often involves multidisciplinary team care.

to R1 was achieved, which, hopefully, with adjuvant chemotherapy and radiotherapy, will delay almost certain death from the tumour.

In conclusion, we describe a rare case of intravenous leiomyosarcomatosis with intracardiac tumour extension that was successfully resected in a combined one step surgery through a midline laparotomy and sternotomy, with circulatory bypass and circulatory arrest. A multidisciplinary team approach was employed in preoperative counselling, surgical planning, operative techniques as well as in postoperative care, to achieve good surgical outcome.

Acknowledgements The authors thank the nurses and operating theatre staff who made this operation a success.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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