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CASE REPORT



## Radical surgery of a recurrent low grade endometrial stromal sarcoma with a comprehensive intravenous growth from ovarian vein to the right ventricle – a case report

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### Introduction

Low grade endometrial stromal sarcoma (LGESS) is a rare malignant disease with an indolent behaviour and propensity for recurrences after long latency period (Leath *et al.* 2007). In some patients LGESS extends directly into vessels reaching the heart, thus increasing the risk for heart complications and pulmonary embolism. Taking into account the high risk for sudden death as well as indolent growth of the tumour with a high 5-year overall survival, the recommended treatment in these patients is early and radical surgery, known to significantly prolong their disease-free survival (Yoon *et al.* 2014, Seagle *et al.* 2017).

Herein we describe a patient with a recurrent LGESS having a comprehensive intraluminal tumour growth, from ovarian vein to the inferior vena cava (IVC) and right ventricle. The patient was surgically treated with a complete resection being accomplished.

### Case report

A 59-year-old patient was admitted with shortness of breath. Her past history included hysterectomy and bilateral adnexectomy for LGESS at the age of 40. Magnetic resonance imaging (MRI) of the pelvis revealed a large retroperitoneal tumour measuring 32 × 47 × 90 mm, with signs of ureteral obstruction. Abdominal and pelvic contrast-enhanced computerised tomography (CT) confirmed the presence of the pelvic tumour and obstruction of the ureter, with the left kidney identified as hypofunctional and atrophic (Figure 1(A)). In addition, large filling defects of the inferior vena cava and renal vein, indicative of thrombosis, were also revealed (Figure 1(A)). The preoperative imaging did not show any enlarged lymph nodes. Dynamic contrast-enhanced heart MRI confirmed the presence of the IVC malignant thrombus extending to the right atrium. Due to the intracardiac tumour


and the risk for acute embolisation and heart failure the patient underwent an immediate surgical resection.

We used a combined sternolaparotomic approach and gynecological and cardiovascular surgical team combined performed the procedure. The retroperitoneal tumour was approached through the median laparotomy and the entire tumour mass adherent to the iliac vessels was separated, without vessel resection. The retroperitoneal tumour extended cranially near the vascular branches of middle colic artery and inferior mesenteric artery, infiltrating them. Complete resection of the tumour was done. Due to the injury of vascular branches supplying left colon and consequential ischaemic changes which were evident, left colectomy had to be performed. The cranial extension of the tumour is shown on Figure 2 (supplemental file). The tumour did not extend to superior mesenteric artery/vein or aorta.

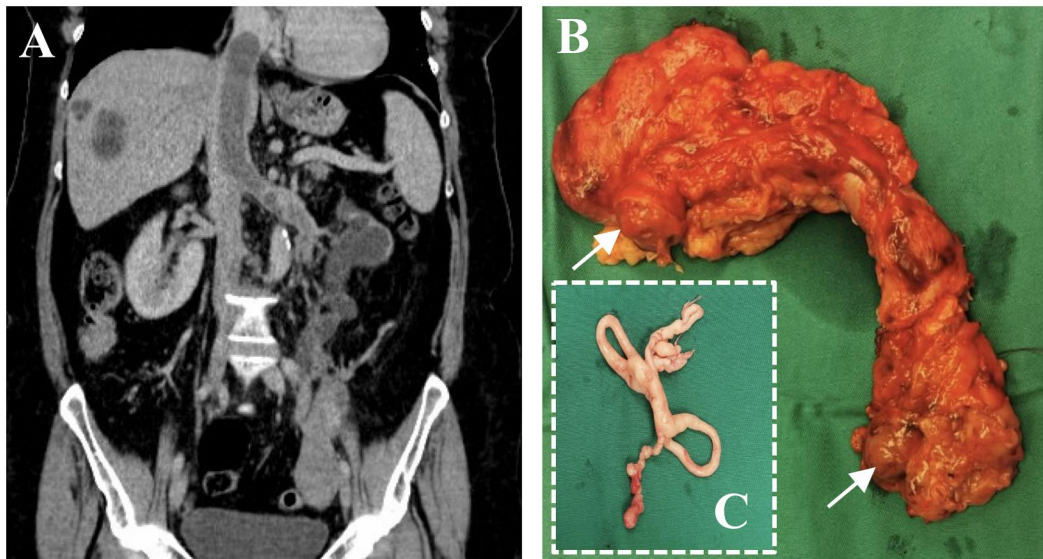
The second step included median sternotomy and instillation of a cardiopulmonary bypass. Aorta, superior vena cava and IVC distally from the pathological process were cannulated. This allowed us to approach the intraluminal tumour through the right atriotomy. The thrombus was mobilised and extracted. At the atrial entry the thrombus firmly adhered so the caval wall had to be resected and sutured. Next, left nephrectomy was performed using a standard procedure with distal resection of the dilated left ovarian vein.

The final histopathologic finding confirmed the recurrence of LGESS with a prominent intravascular part. Macroscopically, retroperitoneal pelvic tumour was a 10 cm large mass, with multiple nodules on cross-section. Microscopically, malignant cells had small nuclei with scant cytoplasm and up to 4 mitosis per 10 high-power fields (HPFs). Immunohistochemically, tumour cells were diffusely positive for CD10, single for p53, while the reaction to caldesmon was negative (Figure 3, supplementary file). Oestrogen and progesterone receptors were positive in 100% of tumour cell nuclei at high staining intensity, and Ki-

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**Figure 1.** (A) Coronal contrast-enhanced CT image of the abdomen and pelvis showing pelvic tumour, atrophic left kidney with dilated ureter, filling defects of IVC and left renal vein, (B) the removed left kidney with a distended left ovarian vein and left renal vein, the vessels are obliterated with tumour visible at the edges due to venous wall retraction (arrows), (C) the cavoatrial smooth malignant thrombus.

67 proliferation index was up to 10%. The intravascular part included a completely obliterated and dilated 3-cm-wide by 13-cm-long left ovarian vein, with the tumour propagating through the left renal vein (Figure 1(B)). As seen in the figure, the perivascular fatty tissue also contained nodular aggregates of malignant cells. The cavoatrial thrombus was a 4 cm large branching mass with a smooth surface (Figure 1(C)), histologically showing areas of hyalinisation and areas of tissue built similar to the pelvic tumour. A part of the lateral atrial wall and IVC were resected ( $4 \times 1.5$  cm) and the reconstruction was done with a continuous suture in two layers (Prolene 4-0). The excised caval wall showed infiltration of malignant cells in the intima. The duration of the operation was 3h and 57 min, the patient received 1980 ml concentrated erythrocytes and 1000 ml of fresh frozen plasma, with the estimated blood loss to be around 2500 mL. On postoperative Day 12, the patient was discharged. The patient received a hormonal adjuvant treatment, megestrol (Megace). During regular check-up, 12 months later, the patient was admitted at our Institution with the suspicion of a pelvic disease relapse.

## Discussion

LGESS is a rare uterine tumour, mostly occurring in women 40–50 years of age. It has a high relapse rate of around 40–50%, especially after a longer time period (Amant *et al.* 2009). Despite its slow growth, LGESS widely permeates the myometrium as a primary tumour and shows tumour plugs in vascular spaces (Nucci 2016). It rarely extends to the major vessels however. In the literature, there are barely more than twenty reports of such cases and none from Croatia. Here we present a patient with a recurrent LGESS having a comprehensive intravascular involvement. Namely, our patient had a heart ultrasound which confirmed the intraventricular propagation of the tumour, which was extracted intraoperatively.

The tumour recurred locally as a large pelvic retroperitoneal mass adherent to the iliac vessels that was successfully peeled from the vascular structures. The intravascular part was extensive requiring resection of the ovarian vein, renal vein and extraction of the cavoatrial tumour with small excision of the cava due to wall invasion. Other published case reports also elucidate this invasive behaviour of LGESS, with one report showing infiltration of aortic outer adventitia (Busuito *et al.* 2012) and another invasion of infrarenal aorta and IVC treated by en bloc resection (Renzulli *et al.* 2009).

Although LGESS is a slow-proliferating nodular tumour, its intravascular growth and invasive behaviour increase the complexity of surgical procedures necessary for oncological clearance. Since there was no visible tumour at the end of operation (R0 resection) our assumption is that the early recurrence in our patient could be due to an aggressive transformation of the tumour into high-grade ESS, which has already been hypothesised (Zou *et al.* 2020). Adequate pre-operative preparation, multidisciplinary approach and a combined sternolaparotomy for complete tumour visualisation are the key components for these patients to be successfully radically operated, which was achieved in our patient.

## Patient consent

Written informed consent for publication of their details was obtained from the patient. Ethics committee approval was unnecessary due to nature of the study.

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