A one-stage approach to the treatment of intravenous leiomyomatosis extending to the right heart

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This report describes the case of a 60-year-old woman with a history of hysterectomy for myomas, totally asymptomatic, with incidental evidence of a pelvic intracaval mass extending to the right atrium. She underwent a staged procedure (sternothomic and abdominal) through a thoracolaparotomic approach in circulatory arrest and deep hypothermia. Using a one-stage surgical approach, we were able to withdraw one portion of the mass from the right atrium and another from the abdominal inferior vena cava, thus minimizing the risk of unexpected venous or atrial wall injury during surgical manipulation. (J Vasc Surg 2010;52:212-5.)

Intravenous leiomyomatosis (IVL) refers to an intravascular benign proliferation of smooth muscle cells originating from the intrauterine venules and reaching the right heart. Although nonmalignant, cases of sudden death and tumor recurrence have been reported.[1-3]

Our literature search retrieved reports of 200 cases, 68 of which described intracardiac extension.[4,5] Single- or two-stage procedures in the approach to intracardiac and abdominal tumors have been proposed.[6,7] Here, we report a case of intracaval leiomyomatosis (ICL) extending to the heart and successfully treated with a single-stage operation with cardiopulmonary bypass (CPB), deep hypothermia, and circulatory arrest.

CASE REPORT

A 60-year-old woman was referred to our department after computed tomography (CT) scan staging for chronic lymphatic leukemia had incidentally detected an intravenous mass extending from the common and internal iliac veins to the right atrium.

The patient had been completely asymptomatic for dyspnea, recurrent palpitation, thoracic pain, syncopal episodes, and had shown no sign of venous hypertension of the lower extremities such as edema, hyperpigmentation, or liposclerosis. Her medical history was significant in that she had undergone a total hysterectomy and bilateral salpingo-oophorectomy for myomas 6 years earlier.

Abdominal ultrasonography and a second, more detailed CT scan demonstrated an intravenous mass, first described as a thrombus (Fig 1A, B, C). Transesophageal echocardiography showed intermittent prolapse of the mass into the tricuspid annulus. Histological evaluation of transjugular and transfemoral biopsies of the intracaval mass confirmed the diagnosis of leiomyomatosis.

Owing to the risk of incarceration in the tricuspid valve and the need for a radical eradication of the mass, the treatment plan called for a combined cardiovascular surgical procedure. The operation entailed a combination of a transsternal and a midline transperitoneal approach, with CPB in deep hypothermia and circulatory arrest.

Through two separate incisions (Fig 2A), the chest was opened, then the abdomen, and the entire infrahepatic vena cava was isolated; both renal veins, the right common iliac artery, and the left common and the external iliac veins were suspended with umbilical tapes. CPB was established by cannulating the right subclavian artery, with venous drainage through the right femoral vein and the superior vena cava. The right atrium was opened, and the upper part of the tumor could be pulled out (Fig 2B). Immediately thereafter, the distal inferior vena cava, the left common, and the first 2 cm of the external iliac vein were incised with a continuous venotomy; the left hypogastric vein, from which the tumor originated and to which it was firmly attached at the vessel wall, was tied. With the use of an endarterectomy spatula, the remaining part of the tumor was mobilized and extracted after gentle retraction (Fig 2C, D, E). Apparently, there were no adherences along the upper path, but if there were, they created no problems.

After completion of the suture of the venotomy, rewarming was started from a minimum temperature of 19°C; the total CPB time was 124 minutes, including a rewarming time of 55 minutes. Circulatory arrest lasted 33 minutes, and blood loss was approximately 1400 mL, of which 1000 mL was collected intraoperatively and 400 mL from the drains during the stay in the intensive care unit (ICU).

The tumor was firm and rubbery, without tumor thrombus or areas of friable tissue (Fig 2F). Histological and immunochemical analyses confirmed the diagnosis of ICL (Fig 3A, B, C, D).

The postoperative course was uneventful; after the first 2 days in the ICU, the patient was discharged from our service on

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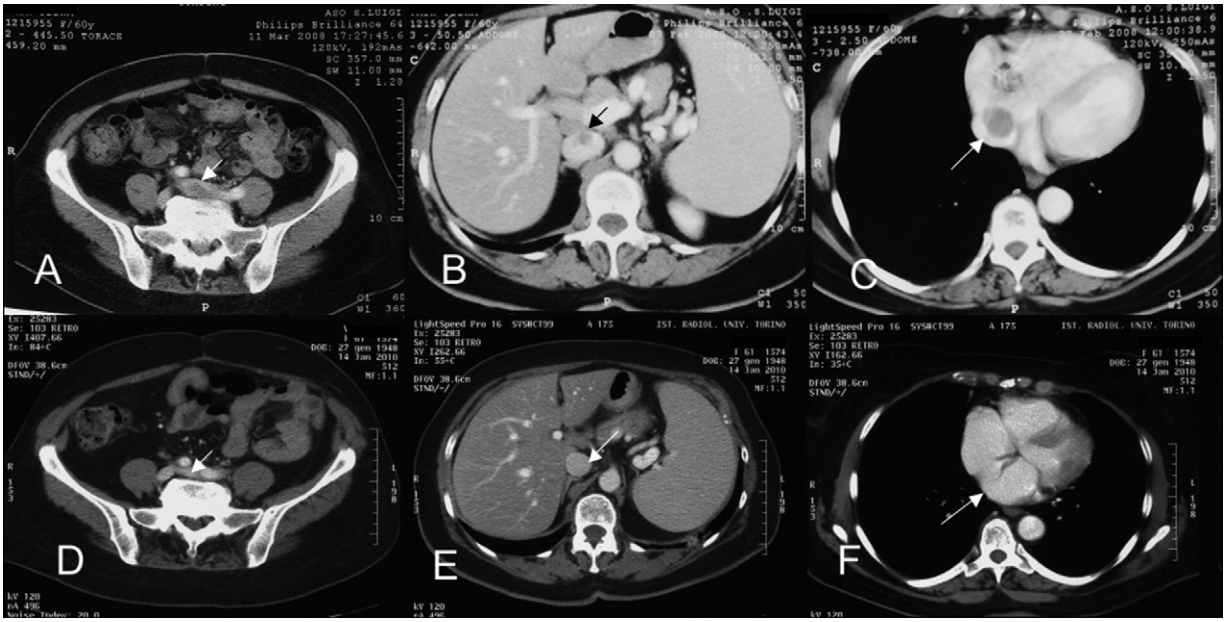

Fig 1. (**A, B, C**) Leiomyoma extending from the external left iliac vein to the right atrium (preoperative). (**D, E, F**) Tumor removed and absence of recurrence (22 months postoperative).

postoperative day 11, under prescription of a daily single-shot dose of low-molecular-weight heparin (LMWH) 5000 IU for 1 month, followed by aspirin 100 mg per day.

At the most recent follow-up visit, about 22 months postoperatively, findings on two control spiral CT scans and two venous peripheral and caval duplex scans were negative for tumor recurrence and thrombotic venous obstruction (Fig 1D, E, F). The patient is being followed up for chronic lymphatic leukemia.

DISCUSSION

Worley et al[8] recently reported their single-institution case series of four operated patients, three of whom were treated with a single-stage approach; in only one of whom radical removal of the tumor was achieved.

In 1982, Ariza et al[9] described the first successful two-stage removal of an intracaval mass, with delayed laparotomy after resection of the intracardiac portion of the tumor. Since then, two surgical techniques have been developed for the removal of ICL extending up into the atrium. One requires single-stage surgery with complete removal of the mass through a simultaneous thoracic and abdominal approach under cardiopulmonary bypass and circulatory arrest with hypothermia, as in our case. The other entails two stages: the first intrathoracic and the second abdominal.[5,6] Both techniques can also be applied in the presence of uterine leiomyoma, from which the concomitant ICL originates.[10]

In their reviews of patients with IVL extending to the heart, Harris and Grella[6,7] described 36 cases treated between 1975 and 1999, 15 of which were treated with the two-stage and eight with the one-stage technique.

We think that, whatever the surgical approach chosen, the aim of treatment should be complete tumor resection to prevent possible long-term recurrence, as described in the literature.[11] The intracardiac portion of the tumor is removed first through a transsternal approach, followed by iliac caval venotomy to extract the tumor remnants. During removal of the intra-atrial portion of the mass, the tricuspid valve can be inspected and eventually repaired.

Notably, attempting to remove the entire caval mass only from the thoracic area may seriously raise the risk of hemorrhage, because of the potential presence of dense adherences along the vascular path, which may result in fatal hemorrhage, as described in the literature.[12,13] Some authors instead have stated that, in principle, these adherences should not exist or, if they do, they will be very loose owing to the characteristics of the tumor, thus allowing its safe removal from the downside without particular risks, possibly with the aid of endovascular techniques and obviating the need for CPB.[14] The one-stage treatment of IVL clearly places a much heavier burden on the patient. If, because of poor condition due to cardiac and pulmonary comorbidities in particular, the patient is ineligible for single-stage surgery, a two-stage operation or perhaps even no operation at all may probably offer the best options.[15] In those cases in which complete tumor removal cannot be achieved for any reason, appropriate hormonal therapy could be recommended following accurate immunohistochemical and receptorial study of the neoplastic tissue.[16]

In our experience, when a two-stage procedure has been planned, the abdominal part of the operation should be done first, because of the much higher probability of encountering dense adherences to the vascular wall at the origin of the tumor, namely, at the inflow point of the hypogastric vein, as found in our case, while the adherences along the upper path were loose and virtually nonexistent,

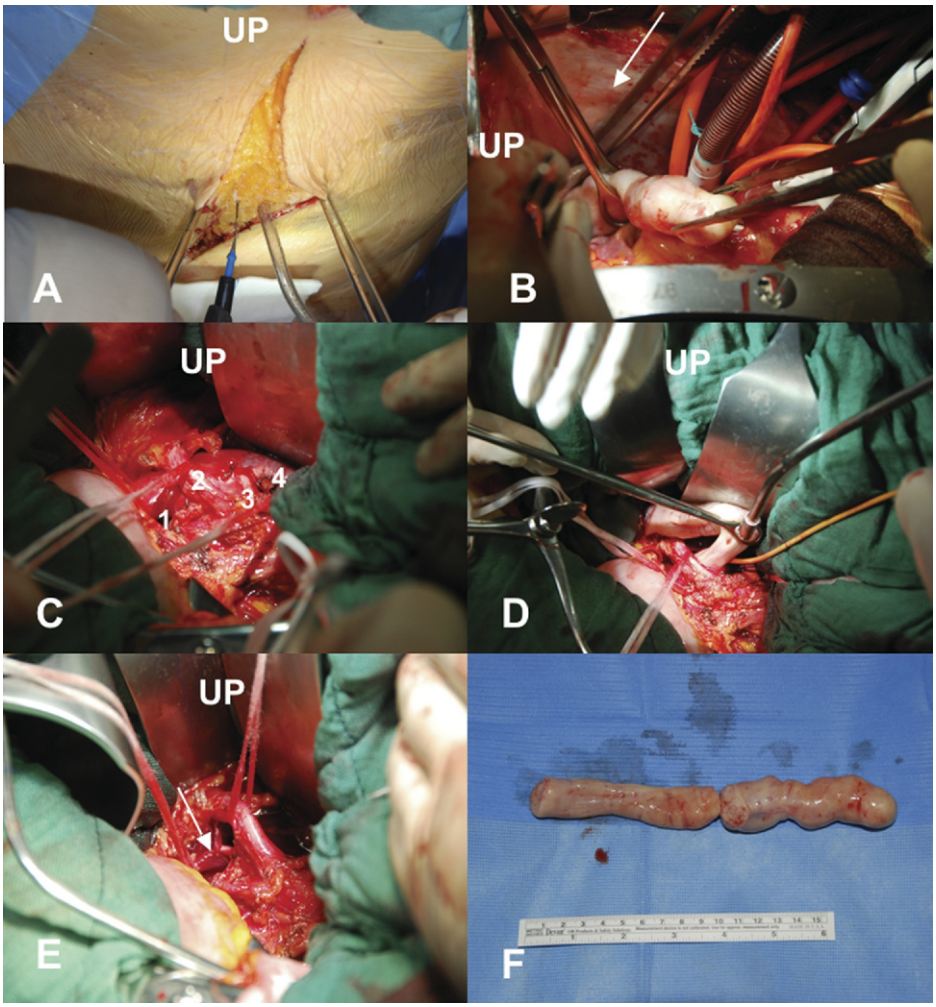

Fig 2. (**A**) Beginning of midline laparotomy and partial view of the operative field, including the distal segment of the sternotomy incision. (**B**) The tumor is pulled out from inside the right atrium and gently stretched, then interrupted as distally as possible. (**C**) The distal vena cava and the first tract of the left common iliac vein (1); the right common iliac artery suspended on umbilical tape (2); the inflow point of the left hypogastric vein (3), and the left external iliac vein (4). (**D**) After venotomy of the distal vena cava, the common iliac vein, and the first segment of the external iliac vein, the tumor is pulled down from the upside and freed from dense adherences arising from the inflow point of the hypogastric vein, which is tied. (**E**) The venotomy is closed with a Prolene 6–0 surjet. (**F**) The tumor is shown after complete removal.

even though we were unable to confirm this before operating.

In our single experience, the one-stage approach proved to be safe and radical, since we were able to manage both the cardiac and abdominal fields simultaneously, minimizing the risk of iatrogenic atrial or caval injury and reducing patient discomfort and the risk of a second operation. This contrasts with reports of other cases in which the risk was higher due to the need to remove an ovarian mass as well,[5] which lengthened the operating time; nonetheless, even in this case, the one-stage approach was suggested.[10]

We agree with those authors who advocate surgery through a single transsternal and laparotomic approach,[7,17] and we feel that this approach can be safely performed also in the presence of an original uterine tumor, following hysterectomy and salpingo-oophorectomy. In our patient, the histological features, past medical history, and cardiac extension of the mass all support the hypothesis for a leiomyomatosis that originated at the pelvic level from the uterine myoma removed years earlier.[1,7,18]

Of note is that no definitive prognosis can be made as to progression of a residual tumoral mass or its recurrence.

CONCLUSIONS

Currently, there is no single surgical technique for treating IVL; both one-stage and two-stage procedures are described. In our case, a clear preoperative diagnosis and the patient's good condition allowed us to approach the ICL by one-stage surgery with CPB and hypothermic

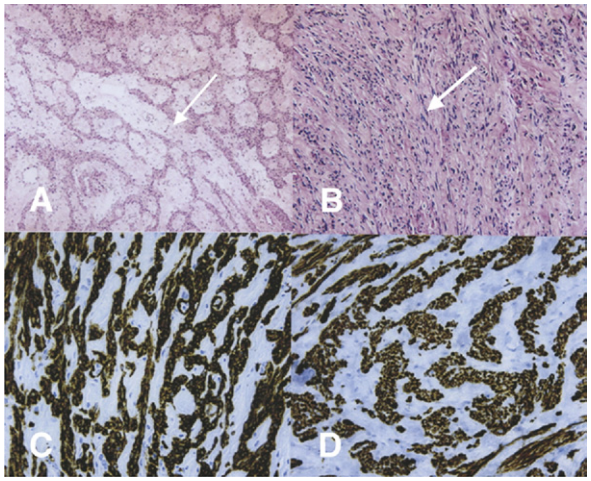

Fig 3. Histological study (**A, B**). Hematoxylin and eosin-stained tissue sections. Muscular fascicles with fusate cells and vessels inside the mass (arrows). Immunochemical study (**C, D**). Actin stain and desmin-stained tissue sections. Bundles of brown muscular cells, which are typical of leiomyomatosis.

circulatory arrest in order to obtain a bloodless operative field.

We think that this surgical strategy, especially in a setting such as ours, where a cardiac and a vascular team can work together, is to be preferred.

Given the lack of a clear understanding of the natural history of this tumor and the relatively high possibility of its recurrence, we recommend a radical strategy in its management.

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