Complete resection of intracardiac leiomyomatosis through an abdominal approach under peripheral cardiopulmonary bypass

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Intracardiac leiomyomatosis (ICL) is a rare benign disease. An ICL is defined as an intravenous leiomyomatosis extending to the heart through the inferior vena cava (IVC). Several surgical strategies have already been established according to the morphology of the ICL.1,2 This case study introduces a strategy of single-stage resection through a laparotomy under peripheral cardiopulmonary bypass (CPB).

CLINICAL SUMMARY
A 62-year-old woman without previous medical illness was seen with anorexia. Computed tomography (Figure 1, A) and echocardiography (Figure 1, B) revealed a mass in the right ovarian vein extending through the IVC into the right ventricle. The surgical team decided to pull out the intracardiac part through cavotomy with the aid of peripheral CPB.

On median laparotomy, the IVC between the level of the renal veins and the level of several centimeters below the ovarian vein was exposed and suspended with umbilical tapes. After heparinization, the extracorporeal circuit was set up with the left femoral artery cannulation with an 18F cannula. Venous drainage was achieved by cannulations of the left femoral vein with a 24F cannula and the right internal jugular vein with a 16F cannula placed through a percutaneous puncture. No chest incision was involved, and the heart continued beating.

After the initiation of CPB under normothermia, a 5-cm longitudinal cavotomy was made at the level of right ovarian vein inlet. The bleeding was controlled by umbilical tapes around the cavotomy and aspirated by 2 intracardiac suction devices. The upper part of the tumor was gently pulled out under surveillance of transesophageal echocardiography (Figure 1, C). The right ovarian vein and the tumor were excised together with a small ellipse of vena cava to which the tumor adhered (Figure 1, D). After suture of the cavotomy, CPB was tapered and protamine was administered. The total hysterectomy and bilateral salpingo-oophorectomy procedure was performed by gynecologist afterward (see details in Video 1).

CPB time was 20 minutes. Operative time was 3.5 hours. The blood loss was merely 100 ml with no blood transfusion. Histological analyses confirmed the diagnosis of ICL. The patient recovered uneventfully and was discharged on the tenth postoperative day. The patient is doing well without recurrence (Figure 1, E) 3 months after surgery.

DISCUSSION
Resections of the intracardiac and intravenous tumors through atriotomy and venotomy in a one-stage or two-stage manner are the common strategies for surgical treatment of ICL1,2; however, patients have to undergo double incisions (ie, thoracotomy and laparotomy). Unlike other intra-IVC tumors, such as the friable renal tumor, ICL is characteristically of high tensile strength. We once hung weights equal to 1 kg on an ICL that had been resected through a sternolaparotomy approach, and the tumor tissue

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was not torn apart by such pulling force (Video 1). The intracardiac part of the tumor can be technically extracted from infradiaphragmatic venotomy if the diameter of it is smaller than the orifice of IVC inlet. Furthermore, the small-diameter tumor means that the tumor lies freely inside the heart chambers and is not likely to adhere to endocardium or the tricuspid valve. For these, extraction of tumor can be achieved by venotomy through a laparotomy alone, through which the hysterectomy and bilateral salpingo-oophorectomy can be concomitantly performed.

Only 10 cases\textsuperscript{1,3-5} out of more than 200 cases of ICL\textsuperscript{2} reported in the literature have had complete resection undertaken through a laparotomy alone. This suggests that the practice of this single-incision strategy is not commonly considered for ICL. It is also notable that no CPB was ever applied in the 10 cases in which the abdominal approach was undertaken.\textsuperscript{1,3-5} As a matter of fact, this procedure often runs the risk of massive blood loss for tumor thrombectomy without the protection of CPB, especially in the case of a dilated tumor larger than the size of IVC, multiple strands of tumor, or adhesions to the IVC which requiring extensive venotomy. Tumor thrombectomy under CPB is reliable without risk of hemodynamic instability and blood loss from venotomy. The avoidance of CPB might be the reason that the application of this single-incision approach is not common. The case reported here demonstrates that CPB by peripheral cannulation without sternotomy plays a facilitative role in this very scenario.

On the basis of our experience, adequate surgical field suction by 2 roller-pump suction devices and proper control of major veins can create a relatively bloodless surgical field. Deep hypothermia and circulatory arrest, which require a sternotomy, thus can be avoided. Protamine should be administered before the pelvic resections to minimize bleeding.

In conclusion, ICL extraction through an infradiaphragmatic venotomy can be improved by peripheral CPB. This strategy is expected to expand the indication of...
single-stage resection through a laparotomy alone for the ICL without intracardiac dilation.

References

EDITORIAL COMMENTARY

Case reporter’s notebook: The who, what, when, where, how, and why of extraction of a benign intracaval tumor

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WHO
A patient who successfully underwent removal of a benign tumor extending into the inferior vena cava (IVC).

WHAT
Complete resection of intracardiac leiomyomatosis via laparotomy and an incision in the IVC under cardiopulmonary bypass with a beating heart and without deep hypothermia and circulatory arrest.

WHEN
Resection of intracaval tumor at the same time and through the same incision as planned hysterectomy and bilateral salpingo-oophorectomy.

WHERE
The entire operation was performed through a laparotomy incision without thoracotomy and with peripheral cardiopulmonary bypass. Arterial cannulation was in the femoral artery with venous cannulation of the femoral and jugular veins. The IVC was isolated and controlled between the renal veins and several centimeters below the ovarian vein that was the entry point of the tumor into the vena cava.

HOW
After laparotomy and isolation of the IVC, peripheral cardiopulmonary bypass was initiated. The heart remained beating throughout the procedure. The distal vena cava was controlled by constriction of the distal umbilical tape encircling the IVC near the ovarian vein. Until it was removed, the tumor thrombus controlled bleeding from the proximal cava and pump suckers were used to maintain a reasonably dry field. After removal of the tumor, the proximal caval tape was tightened to secure retrograde bleeding.