CASE REPORT

Anesthetic Management of a Rare Case of Uterine Leiomyoma with Intravenous and Intracardiac Leiomyomatosis

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Abstract

Background: Intravenous leiomyomatosis (IVL) is an uncommon entity characterized by the growth of benign, smooth muscle tumors within the venous system. Intracardiac extension of this tumor is rare, and very few cases have been reported in the literature.

Case description: We describe a case of IVL with intracardiac extension in a 55-year-old woman, who presented with severe dyspnea, ascites, and lower limb edema. Two-dimensional transthoracic echocardiography and computed tomography showed that the pelvic mass infiltrated the inferior vena cava *via* the iliac vessels and was extending to the right ventricle and pulmonary artery. The patient underwent a one-stage multidisciplinary thoracoabdominal operation with cardiopulmonary-bypass and deep hypothermic circulatory arrest.

Conclusion: Due to the rarity of the present pathology, awareness is widely scarce and diagnosis is often deferred. A correct timely diagnosis, reasonable perioperative plan, and radical excision guarantees favorable outcomes.

Keywords: Anesthesia, Cardiopulmonary bypass, Deep hypothermic circulatory arrest, Intracardiac leiomyomatosis, Intravenous leiomyomatosis.

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Introduction

Intravenous leiomyomatosis (IVL) is a rare histologically benign smooth muscle cell tumor that originates from uterine fibroids and exhibits malignant growth patterns.[1] Usually confined to the pelvis, IVL can occasionally penetrate the ovarian/iliac veins reaching the inferior vena cava, the right heart chambers, and extend to the main pulmonary trunk,[2] resulting in intracardiac leiomyomatosis (ICL) with severe cardiac symptoms and sudden cardiac arrest.[3] Complete surgical removal of thrombus is the treatment of choice. The possibility of massive bleeding and pulmonary embolism is very high, leading to sudden death during manipulation, making perioperative anesthetic management very challenging. We present a case of uterine leiomyoma with IVL extending into the right ventricle and pulmonary trunk, which was successfully removed by median sternolaparotomy under cardiopulmonary bypass with deep hypothermic cardiac arrest (DHCA).

Case Description

A 55-year-old woman with uterine leiomyoma with tumor thrombus extending into the right ventricle was scheduled for thoracoabdominal surgery under cardiopulmonary bypass (CPB).

Our patient was obese, a known case of diabetes mellitus, hypothyroidism, hypertension, bedridden for the past 3 years due to longitudinal extensive transverse myelitis. She presented with severe breathlessness, orthopnea, and bilateral pitting edema in lower extremities. Preoperative evaluation revealed massive ascites, bilateral pleural effusion, a ball-valve thrombus in the RA and RV with extension into the PA, and tricuspid regurgitation. Preoperative CT abdomen delineated that the left internal/common iliac veins and the entire extent of the IVC had arterial/late arterial enhancing tumor thrombus. In addition, multiple tubular and serpiginous enhancing vascular structures representing collaterals were seen in the left hemipelvis and left lumbar region. The uterus was bulky with multiple intramural fibroids and a large left adnexal mass lesion occupying the left hemipelvis, encasing the left distal ureter and left internal iliac vessels (Figs 1 and 2).

The patient was prepared for the surgery after taking adequate consent. Inj Midazolam 0.15 mg/kg IV Inj Ranitidine 50 mg and Inj Metoclopramide 0.15 mg/kg were administered as preanesthetic medications 30 minutes before taking the patient into the theater. Standards monitors were applied according to the American Society of Anesthesiologists and baseline vitals were noted. Due to severe orthopnea, the patient was preoxygenated and intubated in propped-up position with Inj Etomidate 0.3 mg/kg in titrated dose, Inj succinylcholine 2 mg/kg IV using a video laryngoscope and put on mechanical ventilation with pressure control mode. The left internal jugular vein was cannulated with a triple lumen central venous catheter, thus avoiding the risk of dislodging the thrombus in the right atrium. An arterial line was also acquired in the left radial artery. Moreover, transesophageal echocardiography (TEE) was performed during the operation to detect pulmonary embolism and to recognize the position of the thrombus.

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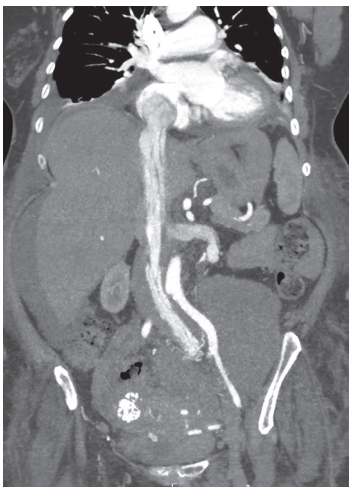

Fig. 1: Coronal postcontrast image of lower thorax and abdomen showing heterogeneously enhancing mass within IVC, seen extending into the right atrium

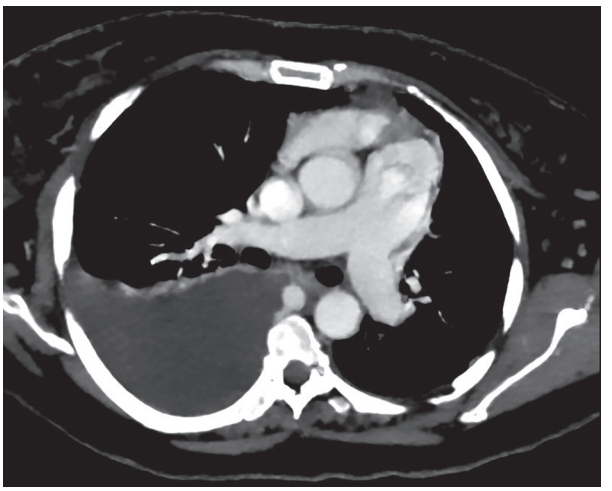

Fig. 2: Axial postcontrast CT section showing main pulmonary trunk with right and left pulmonary arteries with mass extending into the main pulmonary trunk

Rectal and nasopharyngeal temperatures and bispectral index were also monitored intraoperatively.

An infusion of Fentanyl 20 mcg/hr was used as an analgesic intraoperatively and the patient was maintained on oxygen, the air in 1:1 ratio, and sevoflurane along with cisatracurium infusion as a muscle relaxant.

Considering the risk of intraoperative pulmonary embolism, a median sternotomy was performed at the beginning of the operation, to enable us to initiate CPB immediately. Injection Heparin 300 U/kg was administered to achieve and maintain an activated clotting time of >450 seconds and the patient was put on CPB. DHCA was achieved as the patient was cooled to 18°C, over 30–40 minutes using a cooling blanket and ice packs. The temperature gradient between the venous inflow to the CPB circuit and the arterial outflow was maintained at less than 10°C. The patient's head was packed in ice to prevent passive rewarming. Anterograde cerebral perfusion was maintained throughout the duration of circulatory arrest. Blood glucose levels were maintained below 180 mg/dL. Injection thiopentone 5 mg/kg was added to the circuit before achieving DHCA. Arterial blood gases were maintained according to the pH-stat method. After achieving circulatory arrest, the abdominal part of the inferior vena cava (IVC) was exposed by a midline longitudinal incision. Right atriotomy showed thrombus impinging on the tricuspid valve. IVC was opened and the thrombus from the mid-IVC till the right atrium was milked out.

DHCA was maintained for a duration of around 50 minutes during which the thrombus was removed. CPB was reinstituted and rewarming was done slowly over 90 minutes avoiding hyperthermia and maintaining the arterio-venous temperature gradient of less than 10°C. After achieving normothermia protamine was administered and the patient was taken off the CPB. The total pump time was 3 hours.

During dissection of the uterine leiomyoma, the patient bled profusely (total blood loss by the end of the surgery was around 4–5 liters) leading to hypovolemic shock which was supported with infusions of noradrenaline, dobutamine, vasopressin, and transfusion of multiple blood products (15 packed red blood cells, 22 cryoprecipitates, 12 fresh Frozen plasma, 3 single donor platelets). Treatment was guided by serial arterial blood gas analysis, thromboelastography, prothrombin, and activated prothrombin times, INR and fibrinogen.

The surgery lasted for around 12 hours. Postoperatively, the patient was shifted to the intensive care unit (ICU) on a ventilator with noradrenaline infusion ongoing. Histopathological evaluation confirmed the diagnosis of uterine leiomyoma with intravenous leiomyomatosis.

In the ICU, she was weaned and extubated on postoperative day 1 and started on nasogastric feeds. But due to desaturation, she had to be reintubated. Post intubation, her chest X-ray revealed changes suggestive of transfusion-related acute lung injury (TRALI) for which she was treated with intermittent positive pressure ventilation and supportive care. Gradually her condition improved and she was weaned off the ventilator over a week a repeat CT scan showed no signs of tumor thrombus or leiomyoma. The patient is being followed up by her primary physician.

Discussion

Uterine leiomyoma with IVL is an uncommon, histologically benign neoplasm usually confined to the uterine venous system. Rarely it invade the systemic veins reaching the right atrium and ventricle causing right-sided congestive heart failure.

Since the recurrence is known to occur in 1/3rd of cases with incomplete resection, radical surgical removal of the tumor thrombus along with total abdominal hysterectomy and bilateral salpingo-oophorectomy is imperative.[4,5] Complete resection requires a multidisciplinary approach involving general and cardiac surgery in one or two-staged procedures.

In 1974, Mandelbaum et al.[6] reported the successful removal of intravenous leiomyomas involving the right heart.[6] In 1982, Ariza et al.[7] reported the first complete use of staged surgery to remove ICL.[7] The patient in our case underwent a single-stage abdominothoracic operation with a combined sternolaparotomy and cardiopulmonary bypass with deep hypothermic circulatory arrest.

The proximal cardiac tumor thrombus was removed by right atriotomy and incising the IVC. After termination of CPB, pelvic dissection of the uterine mass was started which took 6 hours, leading to major blood loss requiring massive transfusion. Hence, we suggest doing the pelvic dissection while still on the pump so that the blood can be autotransfused.

In the postoperative period, she was electively ventilated overnight while her hemodynamics improved. Though the decision to extubate her on day 1 was taken only after reviewing her vital parameters, she should have been ventilated for at least 72 hours postoperatively, considering the risk of TRALI.

Conclusion

Due to the rarity of the present pathology, awareness is widely scarce and diagnosis is often deferred due to initial nonspecific and subtle clinical manifestations leading to delayed diagnosis in advanced stages when cardiovascular repercussions have already been established.

Thus, anesthesiologists and surgeons should more thoroughly understand IVL to achieve a correct timely diagnosis, prepare a reasonable perioperative plan and perform the rational treatment.

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