CASE REPORT

A multi-disciplinary approach to the management of intra-venous and intra-cardiac leiomymatosis: A case report

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Abstract

We herein report a rare presentation of leiomyoma in a 46-year-old female who presented with complaints of shortness of breath on exertion associated with palpitations for the past one year with a history of irregular menstrual bleeding. Ultrasonography of the abdomen and pelvis revealed an enlarged, distorted uterus with multiple intramural fibroids. A trans-thoracic echocardiography was then performed which showed a large right atrial mass. To investigate further, a contrast enhanced computed tomography was performed covering entire chest and upper abdomen. It showed a large lesion in the right atrium of the heart measuring 6x5cm. The lesion appeared hypo dense and was abutting the tricuspid valve and posterior wall of the Right Atrium. It revealed the Right Atrial lesion extending into the Inferior Vena Cava and lower down into the Common Iliac Confluence, Left Common Iliac and External Iliac Veins. A multi-disciplinary approach was undertaken; the patient underwent a single stage procedure involving a total abdominal hysterectomy with bilateral Salpingo-Oophorectomy followed by excision of the intra-cardiac tumour. Recovery was uneventful and the patient was successfully discharged on the 5th post-operative day. Due to the lack of literature and low incidence of this disease, this case report presents a rare opportunity to define management guidelines for such occurrences in the future.

Keywords: Leiomyomatosis, Intra-cardiac Leiomyomatosis, Intra-venous Leiomyomatosis.

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Introduction

Leiomyoma is one of the most common benign smooth muscle tumour of the uterus.[1] On rare occasions it may grow into the vessels and extend up till the Inferior Vena cava or the heart and hence be referred to as Intra-Venous Leiomyomatosis (IVL) and Intra-Cardiac Leiomyomatosis (ICL) respectively.[2] Management of the disease is based entirely on complete surgical resection of the tumour.[3] However, given its paucity of occurrence, a well-defined guideline dictating a clear surgical plan has not been identified as yet. We present our experience of a patient who underwent surgical resection of the Leiomyomata and was discharged successfully without complications.

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Case Report

A 46-year old female presented with the history of shortness of breath on exertion associated with palpitations, sweating and pedal oedema for the past one year. This case was seen in Cardio thoracic surgery unit of Rehman Medical Institute, Hayatabad, Peshawar on the 13th of November, 2019. She also complained of heavy irregular menstrual bleeding that was relieved by the use of Norethisterone. She was a known case of subclinical hypothyroidism. On physical examination, she was pale with a soft, non-tender abdomen that had a round, smooth, non-tender palpable mass in the hypogastrium. There were left-sided mild crepitations on auscultation of the chest. Rest of the systemic exam was unremarkable.

Ultrasonography of the abdomen and pelvis revealed an enlarged, distorted uterus with multiple intramural fibroids, the largest of which was measuring 8.9x6.1cm. On Colour Doppler Imaging it showed increased vascularity. A trans-thoracic echocardiography was performed, which showed a large right atrial mass that was protruding through the tricuspid valve into the right ventricle. A contrast enhanced computed tomography

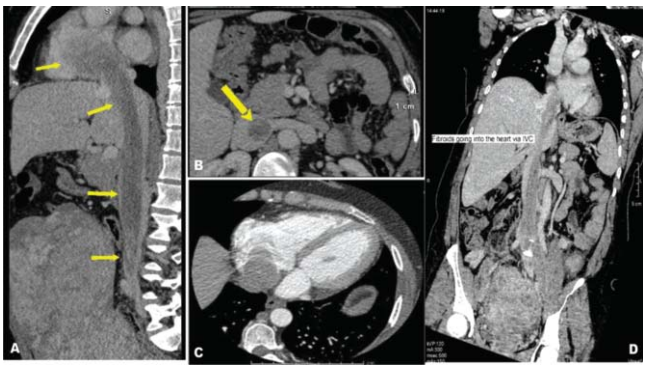

Figure-1: Contrast Enhanced Computed Tomography of the abdomen and chest showing: A) Coronal View: A filling defect in the heart extending through the IVC down to the pelvis. A large Heterogenous Pelvic Mass can also be seen. B) Lesion seen extending into IVC. C) Hypodense lesion seen in the Right Atrium. D) Sagittal view: Large Pelvic Mass seen with direct extension into the IVC and further up into the heart.

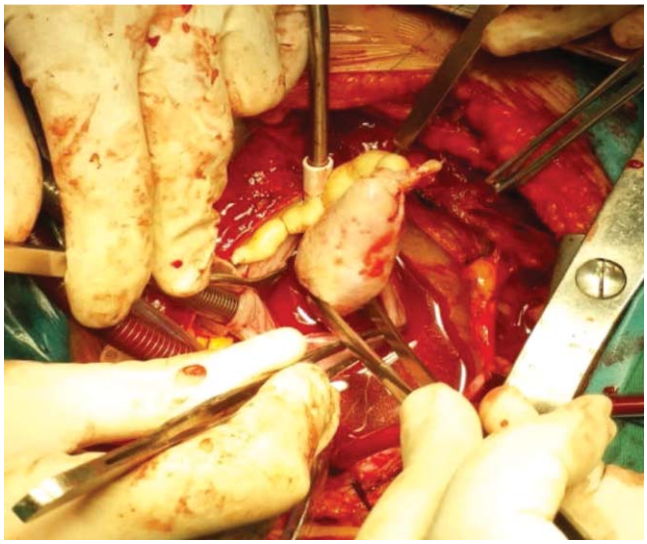

Figure-2: Intra-operative view showing the mass in the right atrium that had entered through the IVC. Pale white object between the forceps.

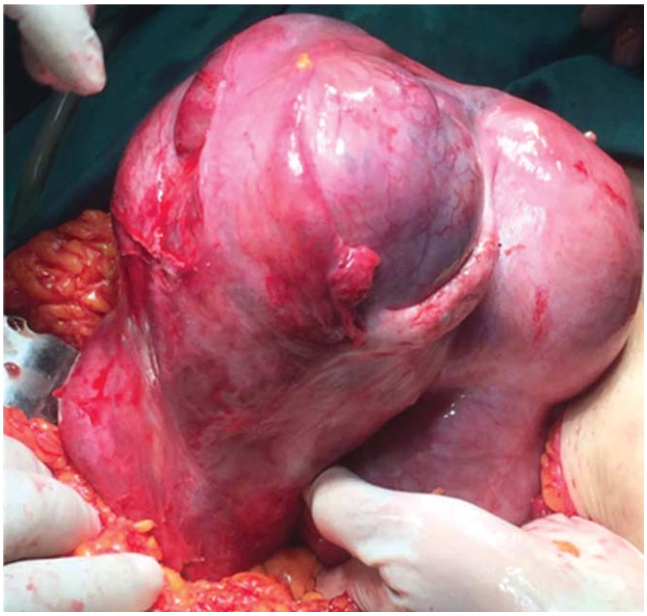

Figure-3: Intra operative image of the uterus and adnexa.

(CT) was performed on 128 slicer Toshiba scanner with computed tomography pulmonary angiogram (CTPA) protocol covering entire chest and upper abdomen. It showed a large lesion in the right atrium of the heart measuring 6x5cm. The lesion appeared hypo dense and was abutting the tricuspid valve and posterior wall of Right Atrium. A delayed venous phase was done for the assessment of IVC covering the lower chest, abdomen and pelvis. It revealed the Right Atrial lesion extending into the Inferior Vena Cava (IVC) and lower down into the Common Iliac Confluence, Left Common Iliac and External Iliac Veins. Its intravenous component had a heterogenous density on venous phase and was not apparently involving any of the venous wall.

A large hyper enhancing 23.5x22x9.5cm uterine mass was seen, representing fibroids involving the fundus and body, with numerous internal dilated vessels and necrotic areas, dilated bilateral ovarian veins were seen and few filling defects were also noted in some of the branches of the Left Gonadal, Left External and Common Iliac Veins. Possibility of tumour thrombus/intravascular Leiomyomatosis was considered which had extended up to the heart. A multidisciplinary meeting was conducted and a presumptive diagnosis of Intracardiac Leiomyomatosis was made. It was decided to proceed with a single stage procedure involving a total abdominal hysterectomy with bilateral Salpingo-Ooporectomy, followed by the excision of the intra-cardiac tumour. Initially, a cystoscopy-guided stenting of the bilateral ureters was performed to avoid iatrogenic injury to the ureters followed by Total Abdominal Hysterectomy by the Gynaecologist and General Surgeon via a mid-line laparotomy incision. An enlarged bulky uterus with multiple adhesions to the ascending colon, descending colon and urinary bladder was dissected and removed along with the bilateral ovaries and fallopian tubes. This was followed by a midline sternotomy. After pericardiotomy, aortic cannulation was done in the ascending aorta. Venous cannula was passed in to the Superior Vena Cava. A Left atrial vent was placed through the right superior pulmonary vein. The patient was cooled to 25°C. Under cardioplegic arrest, the Right Atrium was opened and a blood-less field was maintained using two cardiotomy suctions. Using gentle traction a long, pale, tubular, rubbery mass measuring 35cm in length was pulled from the Inferior Vena Cava. A little manipulation was required from the abdomen as well to free the distal end of the mass from the left ovarian vein. The patient was weaned of cardiopulmonary bypass successfully and uneventfully. The mass was

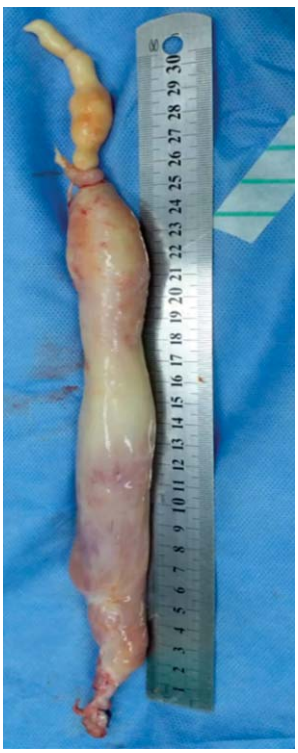

Figure-4: The Leioymyoma in to-to measuring approximately 30cm in length.

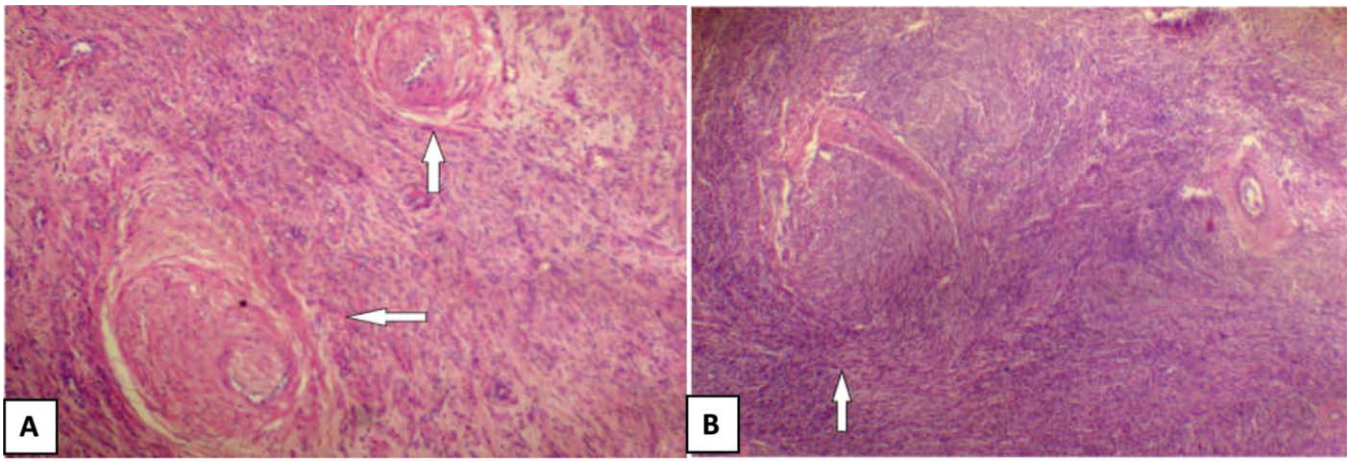

Figure-5: Uterine leiomyoma seen as fasiculated and packed smooth muscle cells arranged in orderly intersecting fascicles. Intravenous leiomyomatosis seen on cut section with the characteristic whorl-like appearance composed of smooth muscle cells separated by variable amounts of fibrous connective tissue.

sent for histopathology which confirmed our diagnosis of leiomyomatosis.

The patient was shifted to the Intensive Care Unit (ICU) and was extubated the same day. She was shifted out of the ICU on day 3 of the surgery and was discharged home on day 5 of surgery.

Discussion

Leiomyoma is a benign smooth muscle tumour most commonly found in the uterus of pre-menopausal women.[4] Rarely, the tumour can exhibit anatomically malignant characteristics by growing into the pelvic veins whereby it is called Intra-Vascular Leiomyomatosis.[5] In these circumstances, a few of these tumours can even grow further up in the venous channel and enter the cardiac chambers, and will then be referred to as Intra-Cardiac Leiomyomatosis.[2] Intra-cardiac and intra-venous leiomyomatosis is a rare condition with only a few hundred reported cases worldwide. The first reported case of an intra-cardiac leiomyoma was by Durck in the year 1907.[6]

Patients of leiomyoma can present with a wide spectrum of symptoms ranging from plain uterine symptoms of irregular and/or heavy menstrual bleeding[3] along with abdominal heaviness and discomfort[7,8] to symptoms of right heart failure like shortness of breath, pedal oedema, ascites, or in some circumstances, with embolic phenomenon in the form of pulmonary embolism.[1,5] Our patient had distinct heavy and irregular menstrual bleeding that lead to the diagnosis of a uterine leiomyoma which is more commonly, known as Fibroid Uterus. A review of the case reports show that a lot of these patients have a previous history of subtotal or total hysterectomy of the uterus for a leiomyoma.[2,3] The intra-cardiac or intra-vascular leiomyoma is later diagnosed on follow-up with a median time between hysterectomy and diagnosis of IVL of 4 years as reported by Teo YM et al.[9]

The first diagnostic evaluation is usually initiated by an ultrasound of the abdomen and pelvis which, in most cases reveals a pelvic tumour extending into the pelvic veins.[4] Symptoms of shortness of breath or pedal oedema require a Trans-Thoracic Echocardiography (Echo) which will show a mass arising from the inferior vena cava and floating in the right atrium that may or may not extend into the right ventricle during the cardiac motion, causing any level of tricuspid stenosis or regurgitation.[1,4] Further evaluation is done using a Contrast Enhanced Computed Tomography (CECT) of the abdomen and thorax.[1,4,5] Magnetic Resonance Imaging provides better differentiation between soft tissues and is the preferred diagnostic modality for viewing soft tissue tumours,[5] but because of its higher cost and longer time consumption, a CT scan may be done instead. In our case, the ultrasound showed an enlarged uterus and bilateral ovaries with multiple masses extending into the left internal iliac vein, the common iliac vein, the inferior vena cava and finally the right atrium as seen on the Echo. The CT scan confirmed that the mass did not have any attachments or pedicles in the venous system or the heart and seemed to be floating freely in the right atrium and inferior vena cava. A presumptive diagnosis of Leiomyomatosis was made based on the history and imaging.

Treatment of intravenous/intracardiac leiomyomatosis is complete surgical resection to avoid recurrence.[1] Surgical approach varies between a single stage versus two stage surgical resection, the decision of which is based on the clinical condition and history of the patient. M.B. Schito et al[1] reported a two stage procedure because of the

increased on-pump time during resection of the intra-cardiac and intra-vascular portion of the leiomyoma. The pelvic tumour was resected in a second operation 7 days later. Similarly, a case series by Hanjang Zeng et al.[5] described an elective two stage resection almost 3 months apart. The authors of this case report preferred a single stage procedure given the clinical condition of the patient.

The intra-cardiac resection can be done under a number of different techniques. We preferred using a single venous cannula in the superior vena cava and an arterial cannula in the ascending aorta at severe hypothermia of 25°C using cardioplegic arrest. The patient was maintained at severe hypothermia to be prepared for the possible need for Deep Hypothermic Circulatory Arrest. This is the only case that we know of that used only a single venous cannula in the superior vena cava. Jonathan D. Price et al. in their case report use of a femoral venous cannula and a superior vena cava cannula for venous return[3]. In addition, a deep hypothermic circulatory arrest can be used to ensure a blood-less field. During our surgery, we snugged the superior venous cannula and ensured a bloodless field via 2 x cardiotomy suctions. The authors of this case report did not use an extra femoral venous cannula because it was felt that due to the large mass almost occluding the inferior vena cava there would not be much return into the femoral venous cannula. Secondly, during procedure the field was bloodless enough with a single SVC cannula that further cemented our belief that a second femoral venous cannula was not required.

There are no guidelines that recommend the follow-up of these patients. However, Yu X et al. have reported a recurrence rate of up to 31%.[10] In our center, we do a routine follow up at one week, one month, 3 months and then 6 months intervals.

Conclusion

Intra-venous and intra-cardiac leiomyomatosis is a rare condition that should be suspected in female patients with a concurrent pelvic and intra-cardiac mass. The condition can be fully and effectively managed by surgical resection that may be done as a single staged procedure if the clinical condition allows.

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Consent: Prior consent was taken from the patient and family for publishing this case report including for pictures that have been used.

Conflict of Interest: We disclose that the person who has signed our ethical review statement is also the co-author of the same manuscript.

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