

# Subserosal Leiomyoma with Intravenous Leiomyomatosis Extending into Inferior Vena Cava and Right-sided Cardiac Chambers

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## ABSTRACT

Leiomyoma (fibroid) is the most common benign tumour of the uterus in reproductive age. Its treatment is easy and employs a surgical resection, usually without complications. However, it may show an atypical behaviour and result in intravenous leiomyomatosis (IVL) extending into the regional and systemic veins, thus reaching the heart; and may extend into the pulmonary arteries. The symptoms of IVL are non-specific, resulting in underdiagnosis and misdiagnosis of the disease entity and so the condition is underreported. Cross sectional imaging plays an important role in correct diagnosis of the condition, resulting in reduced morbidity and mortality of the patient and saving surgeons from unwanted complications. It typically originates from the uterus and may involve ovarian/uterine veins. We report a 45-year female who presented with right hypochondrial pain for which she was diagnosed as cholelithiasis. She was also diagnosed with a left adnexal mass on ultrasonography (USG) for which she underwent a CECT abdomen and pelvis and was diagnosed as a case of subserosal fibroid with IVL extending into left gonadal vein, and inferior vena cava (IVC). Intracardiac extension was confirmed on 2-D echocardiography. This is first reported case of IVL from Balochistan, Pakistan.

**Key Words:** *Leiomyoma, Intravenous leiomyomatosis, Intracardiac extension, Inferior vena cava.*

## INTRODUCTION

Leiomyomas are the commonest benign tumors of uterus in reproductive age. Uncommonly, these may show atypical behaviour such as intravenous leiomyomatosis (IVL). It is noted to show benign intravascular proliferation without invasion into the tissue.<sup>1</sup> Although IVL is commonly limited to the pelvic organs, it may show extension into the pelvic vessels, inferior vena cava (IVC), right sided cardiac chambers, right pulmonary artery; hence, it complicates the case.<sup>2</sup>

Other atypical presentations include peritoneal extension, metastasis to distant organs, leiomyomas reaching beyond peritoneum and parasitic leiomyomas.<sup>3</sup>

Extrauterine involvement and intracardiac extension is noted in approximately 30% and 10% of the cases, respectively.<sup>1</sup> According to Correia *et al.* and Pena *et al.*, very few cases, 300 of IVL extending to heart have been reported in the English literature.<sup>3,4</sup>

According to Du *et al.*, recurrence was related to the surgical approach of the disease.<sup>5</sup> Women undergoing hysterectomy with bilateral salpingo-oophorectomy had less recurrence than women who had hysterectomy alone or just myomectomy.

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*Received: December 19, 2018; Revised: February 22, 2019;*

*Accepted: February 22, 2019*

## CASE REPORT

A 45-year female presented with right hypochondrial pain. She underwent an USG of abdomen in which she was diagnosed as a case of cholelithiasis. USG further reported a complex left adnexal mass for which she was suggested a contrast enhanced CT (CECT) scan of abdomen and pelvis by the gynaecologist. The patient's medical history included a myomectomy few years back. However, no surgical notes/past records were available. CT scan showed a lobulated heterogeneously enhancing mass (13.2 X 8.8 X 8.4 cm) in left hemipelvis, which was in continuity with fundus of uterus (Figures 1a and 1b). Prominent vessels were seen extending from the uterine fundus into the mass lesion (bridging vessel sign). Multiple prominent vessels were seen in the left postero-lateral aspect of the mass lesion, where a prominent dilated left ovarian vein was also seen extending into the posterior aspect of mass, likely representing parasitic flow of the lesion from left gonadal vein. Few linear filling defects were noted in the left ovarian vein (Figure 2a), where it was in continuity with the bunch of vessels in the mass. Few linear filling defects are also noted in the intrahepatic part of IVC (Figure 2b). The mass was extending upto the anterior abdominal wall and posteriorly in close proximity to the left external iliac artery and left psoas muscle. Along its anterior and right lateral aspects, it was abutting small bowel loops. Inferiorly, it was in close vicinity to dome of urinary bladder. Right ovary was normal. Left ovary could not be separately seen. Gall stones were noted. Rest of the abdominal viscera were normal. As the lesion was extending into the IVC, it was suspected that mass might

extend into the right sided cardiac chambers, too. Therefore, she was further advised an echocardiography, which showed echogenic mass extending from IVC into the right atrium and prolapsing into the right ventricle during systole. Mild tricuspid regurgitation was also noted. It was not adherent / originating from the cardiac walls and possibility (after knowing the history) was raised of intracaval and cardiac thrombus / leiomyoma. A one-stage combined multidisciplinary approach, including department of gynaecology, cardiac surgery

and vascular surgery was contemplated; but was not feasible in the setup. She was started on low molecular weight heparin (clexane, enoxaparin).

During its administration, the intracardiac lesion size was monitored by echocardiography, which, however, showed no reduction in the size of the lesion. Patient was operated upon to remove the cardiac mass. The mass had no attachment with cardiac walls. Patient expired few hours after surgery due to massive pulmonary thromboembolism / tumor embolism. The histopathology showed elongated and cigar shaped cells (Figure 3), staining positively for desmin and negative for ki 67, consistent with radiological diagnosis of leiomyoma.

### DISCUSSION

The pathogenesis of IVL remains unclear, however, different theories are proposed suggesting its origin from the venous wall or leiomyoma invading into the uterine vein.<sup>1</sup> As detected by echocardiography, the intra-vascular and intra-atrial tumors were freely movable within the vascular and heart spaces having no attachment to adjacent structures.<sup>1</sup> In this case, it was not adherent to cardiac walls either.

As it extends into the cardiac chambers, it may result in variable cardiac symptoms ranging from fainting to sudden death.<sup>6</sup> However, in this patient, there were no cardiac symptoms and the diagnosis of IVL was made incidentally.

Birch-Hirschfeld presented first case of IVL in 1896,<sup>7</sup> and Durck presented first case of intracardiac extension of IVL in 1907.<sup>8</sup> IVL with intracaval and intracardiac extension has rarely been reported in literature.<sup>9</sup>

Although IVL is a benign entity, it may show a malignant behaviour, due to its growth into the systemic vessels and extension into cardiac chambers and pulmonary vasculature.<sup>10</sup> In this case, a CECT chest was also done, which showed no extension into pulmonary vasculature.

A study reported 52 cases diagnosed with IVL from year 2000 to 2015, in which majority of cases were diagnosed intraoperatively.<sup>9</sup> In this case, diagnosis of IVL was made well before surgery.

According to Du *et al.*, hysterectomy and unilateral salpingo-oophorectomy is an option for women whose family is complete,<sup>5</sup> but when there is evidence of a tumor in the extrauterine veins, hysterectomy, bilateral salpingo-oophorectomy, removal of the extrauterine tumor with high ligation of ovarian veins and arteries and placement of filter into inferior vena cava to prevent embolism is warranted. Unfortunately our patient could not have the privilege of multidisciplinary approach.

In conclusion, IVL is a rare condition that can result in serious complications. Early diagnosis can lead to better patient outcome and less complications on the part of surgeons.

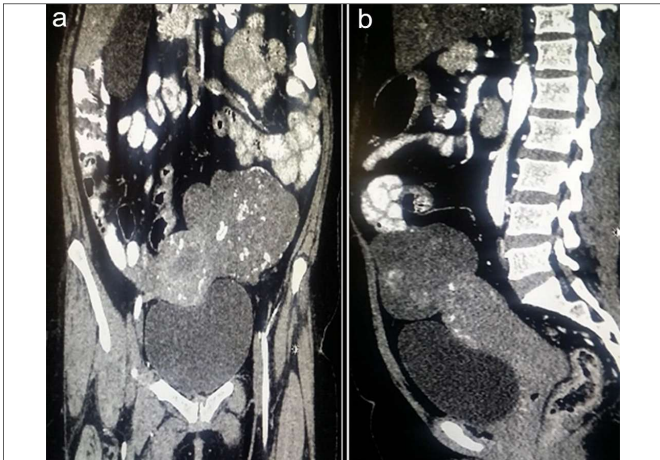


Figure 1 (a,b): Large enhancing mass lesion in left adnexa which is inseparable from uterine fundus. Few prominent vessels are seen within the mass.

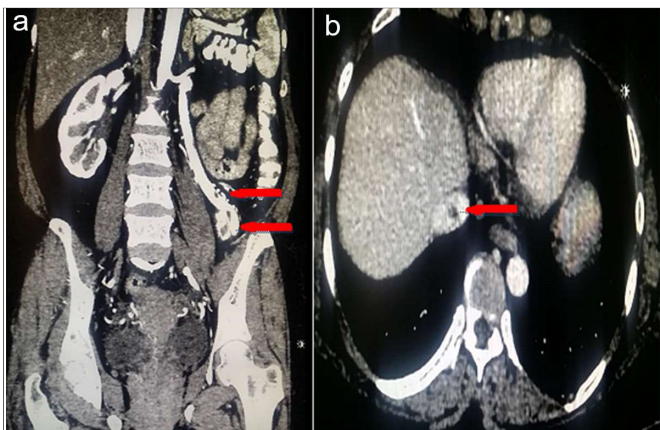


Figure 2 (a,b): Dilated left gonadal vein extending to the mass lesion with few internal hypodense filling defects (2a). Intracaval filling defect is also noted in the intrahepatic part of IVC.

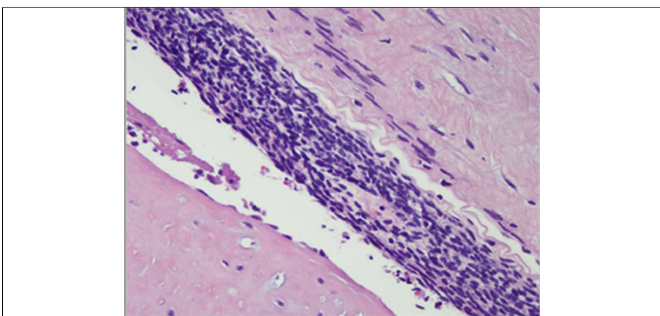


Figure 3: High-power view showing bland spindle cells with elongated and cigar shaped nuclei consistent with leiomyoma.

The case indicates that although leiomyoma is a benign entity, it should be investigated as a systemic disease if it presents as a soft mass in systemic veins, as it has the ability to reach heart and pulmonary vessels leading to serious complications. Secondly, a multidisciplinary approach is very important to make a timely diagnosis and better management.

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