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Case Report

Intravascular leiomyomatosis associated with uterine STUMP extending into the right atrium: a rare case report

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ABSTRACT

Intravascular leiomyomatosis (IVL) is a rare benign smooth muscle tumor that originates from the uterus and proliferates within venous channels, occasionally extending into the heart. Smooth muscle tumour of uncertain malignant potential (STUMP) represents a borderline uterine neoplasm with unpredictable biological behavior. The coexistence of IVL and STUMP is exceptionally rare, posing diagnostic and therapeutic challenges. We report the case of a 52-year-old perimenopausal woman who presented with abdominal distension and bilateral lower limb pain for six months. Imaging revealed a large uterine mass with a thrombus extending from the pelvic and gonadal veins into the inferior vena cava (IVC) and right atrium. The patient underwent staging exploratory laparotomy with total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic tumour excision, venotomy with thrombus extraction, and lymphadenectomy. Histopathology confirmed a spindle cell neoplasm consistent with a STUMP associated with intravascular leiomyomatosis. This case highlights the rare coexistence of STUMP and IVL with intracardiac extension. Comprehensive preoperative imaging, multidisciplinary surgical planning, and complete tumor resection are crucial to prevent recurrence and ensure favorable outcomes.

Keywords: Intravascular leiomyomatosis, Smooth muscle tumour of uncertain malignant potential, Right atrial thrombus, Inferior vena cava thrombus

INTRODUCTION

Uterine smooth muscle tumours have, historically, been classified into benign leiomyomas and malignant leiomyosarcomas according to the degree of cytological atypia, mitotic activity, and other molecular tissue markers.[1] However, there is a spectrum of borderline tumours, including variants of mitotically active, cellular, and atypical leiomyomas, as well as smooth muscle tumours of uncertain malignant potential (STUMP).[2] This term was introduced by Kempson in 1973 and, according to the WHO, should be used for fibroids that cannot be unequivocally and histologically diagnosed as being benign or malignant.[3,4] Most reports of uterine STUMP have found a similar age of diagnosis to that of fibroids or sarcomas, and the median age of this is between 40 to 50 years.[5] Compared to sarcomas, they have a better prognosis, but their biological potential is uncertain, as recurrence and metastases are possible until many years later. Intravascular leiomyomatosis (IVL) is a rare benign condition in which a leiomyoma, propagates through the pelvic venous system into the inferior vena cava (IVC), and occasionally reaching the heart. Despite its low incidence and benign nature, IVL can lead to life-threatening obstructions in the right heart's outflow tract, potentially resulting in sudden death. It was first described by Birch-Hirschfeld in 1896 and is exceptionally rare with fewer than 300 reported cases in the literature. Its

pathogenesis remains uncertain, with two proposed theories: one suggesting IVL originates from uterine leiomyomas infiltrating venous walls and the other proposing it arises from smooth muscle cells within uterine veins.[6] Most of the reported cases have been seen in association with benign uterine leiomyomas and the coexistence of STUMP and IVL is exceptionally rare, with only less than 10 cases reported worldwide. In this article we present such a rare clinical case of intravascular leiomyomatosis associated with STUMP uterus, offering insights into clinical manifestations, diagnostic approaches, and treatment modalities.

CASE REPORT

A 52-year-old perimenopausal woman presented with abdominal discomfort, distension, associated with bilateral lower limb pain for the last six months.

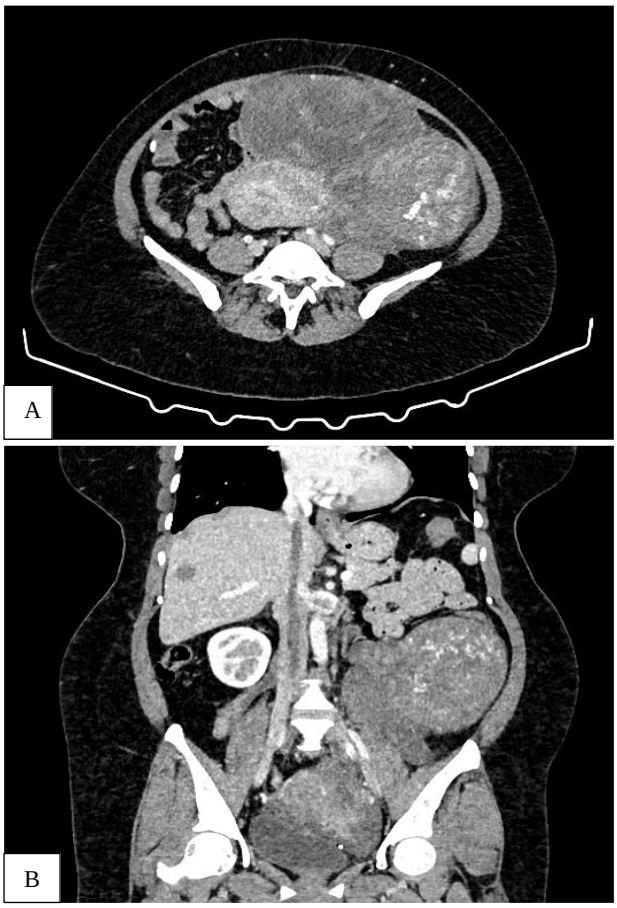

Figure 1(A and B): Large abdomino-pelvic mass predominantly involving the left lumbar region. IVL extending into the right atrium seen in sagittal images.

Examination revealed a firm, irregular, non-tender abdominopelvic mass of 28-week size. Ultrasound revealed an enlarged uterus with multiple fibroids, and MRI showed a large well defined lobulated pelvic-abdominal mass arising from the uterus associated with a partial filling defect in the pelvic and gonadal veins extending into the IVC and up to right atrium. A CECT abdomen with CT angiogram was done which confirmed non-enhancing thrombus within IVC extending to left renal vein and into right atrium (Figure 1). Serum tumor markers were normal (CA 125 - 11.4IU/ml, CEA - 2.2 ng/ml, AFP - 1.04ng/ml, LDH - 210IU/l, Beta HCG<2 mIU/ml, S. Chromogranin A 9.6 ng/ml). USG guided biopsy was taken from the lesion and was suggestive of spindle cell neoplasm showing SMA and Desmin positivity, and low KI-67 index, consistent with a smooth muscle tumor. Intraoperatively there was a large abdominopelvic mass measuring 25×25 cm predominantly occupying the left parametrium along the broad ligament abutting the sigmoid mesentery, upper rectum and extending to the left mesorectum and para cervix (Figure 2).

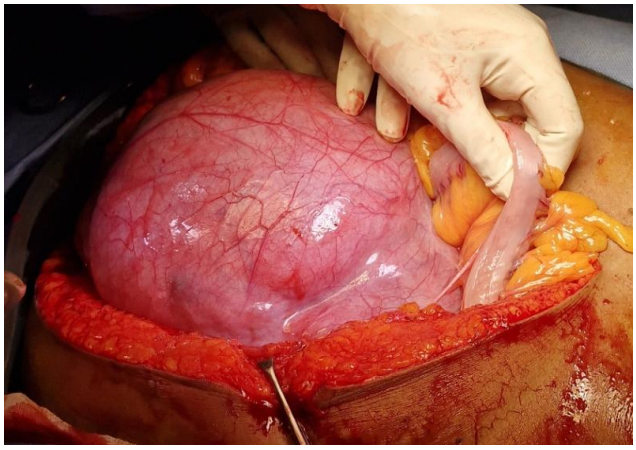

Figure 2: Abdominopelvic mass getting delivered through midline laparotomy.

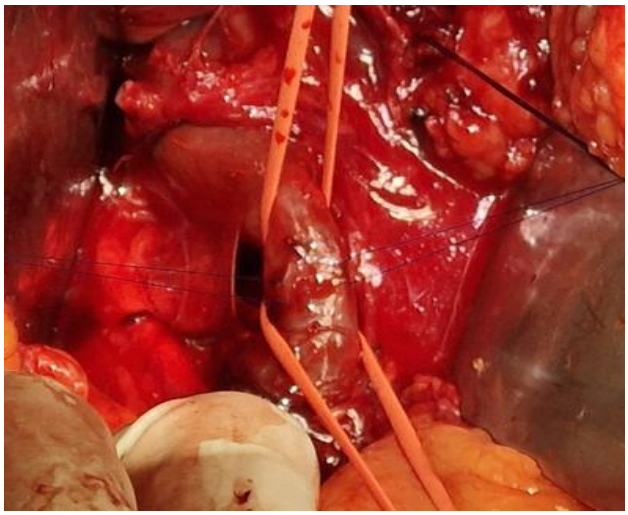

Figure 3: Venotomy being prepared in the anterior wall of infrarenal IVC.

A thrombus was noted in the left ovarian vein extending into the left renal vein and IVC which then was distally extending into the left common iliac vein, and proximally up to the right atrium. She underwent pelvic tumor excision with total abdominal hysterectomy and bilateral

salpingo-oophorectomy. The tumor was dissected off from the left pelvic side wall, left parametrium and left meso rectal region and shaved off from the adhesions with upper rectum and urinary bladder. A 5 cm long venotomy was done on IVC anteriorly at infrahilar region (Figure 3). The thrombus is milked out and extracted superiorly from the right atrium and inferiorly from iliac veins (Figure 4). The venotomy was then repaired using 4-0 prolene (Figure 5).

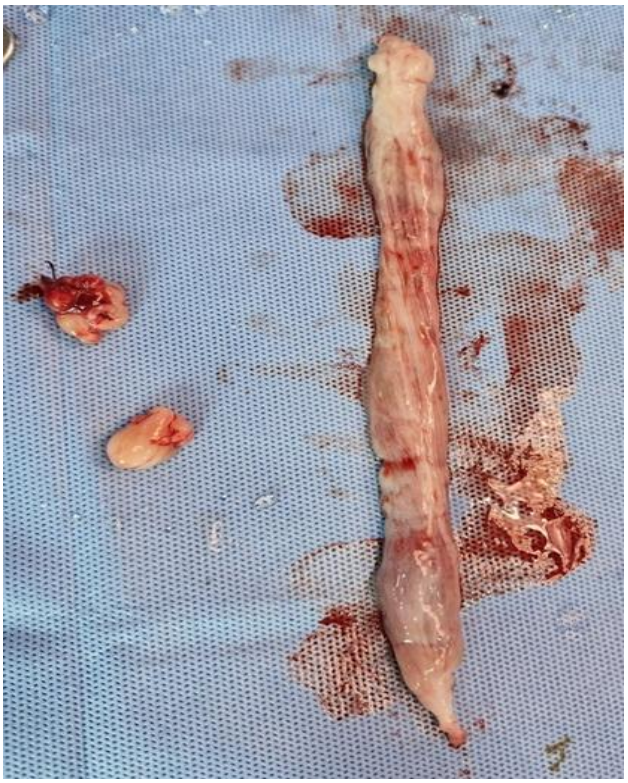

Figure 4: IVL component of the tumor extending up to right atrium.

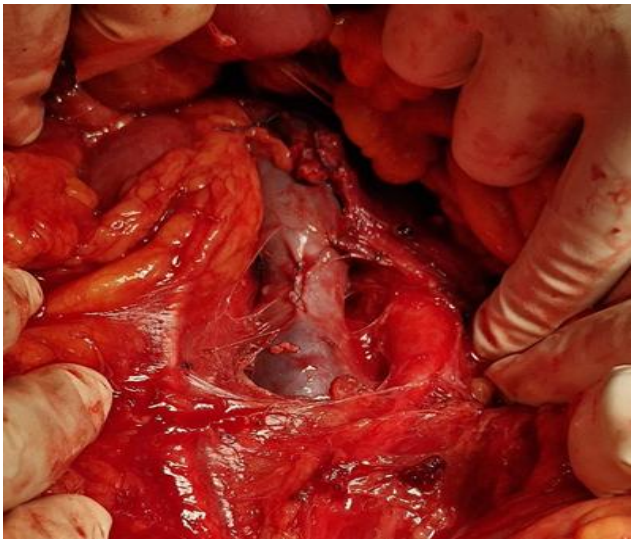

Figure 5: IVC venotomy after repair.

Omentectomy, Paraaortic and pelvic lymphadenectomy were also done. The postoperative period was uneventful and the patient was discharged on postoperative day 10. Histopathological examination of the pelvic mass showed an encapsulated lesion composed of cells arranged as diffuse sheets and fascicles with individual neoplastic cells showing spindle and focal epithelioid morphology.

Numerous thin walled to thick walled ectatic vascular channels are seen scattered. Focal areas show moderate atypia. Sections from the vena cava thrombus also showed areas of smooth muscle proliferation and edema with no definite cytologic atypia. Immunohistochemistry evaluation was also done for confirmation (SM, Desmin - diffuse strong cytoplasmic positivity; CK, CD34, CD10, Melan A, HMB 45, S-100 - negative; KI67 <5%) (Figure 6). Hence a histological diagnosis of smooth muscle tumour of STUMP associated with IVL was made.

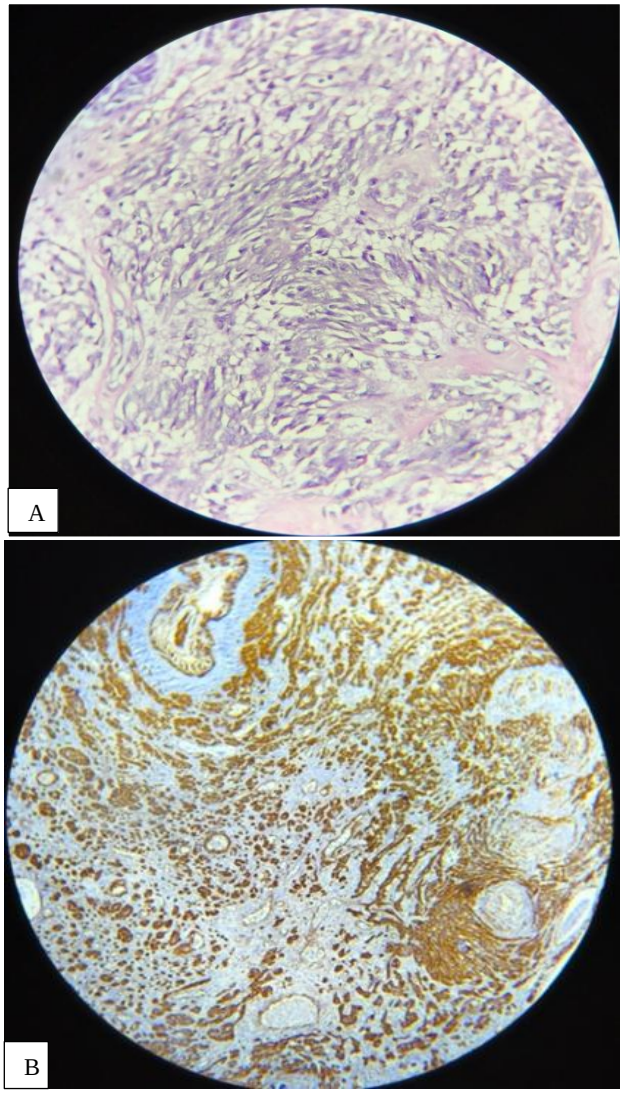

Figure 6 (A and B): H and E: Spindle shaped cells with elongated vesicular nuclei and moderate amount of pale eosinophilic cytoplasm; diffuse staining with H-caldesmon.

DISCUSSION

IVL is an exceptionally rare condition that poses significant diagnostic challenges due to its nonspecific clinical manifestations and low incidence. It primarily affects women of reproductive age, although postmenopausal cases have also been reported.[7] The clinical presentation ranges from asymptomatic to severe, including palpitations and features of right heart failure such as dyspnea, ascites, jugular venous distension, lower limb edema, syncope, and, in rare cases, sudden death.[8] Tumor spread typically follows two main venous pathways: the first via the ovarian vein directly into the IVC or through the left renal vein when originating from the left adnexa—and the second through the internal iliac venous system, ultimately draining into the IVC via the common iliac vein.[9] When IVL extends into the cardiac chambers, the condition is termed intracardiac leiomyomatosis (ICL), a particularly uncommon manifestation. Accurate preoperative evaluation and staging rely on multimodal imaging approaches, including echocardiography (transthoracic or transesophageal), venography, abdominal and pelvic computed tomography (CT), magnetic resonance venography, and CT angiography (CTA). Contrast-enhanced CT and magnetic resonance imaging (MRI) are instrumental in delineating the tumor's origin and tracking its course, even in patients with prior hysterectomy. Some authors suggest that contrast CT provides superior visualization compared to MRI for defining the tumor's full extent.[10] The differential diagnosis includes thrombus, atrial myxoma, and intravascular extensions of malignant neoplasms such as renal cell carcinoma, Wilms' tumor, and adrenal carcinoma. IVL should be suspected in women with a history of uterine leiomyoma or hysterectomy who present with a right-sided cardiac mass continuous with the IVC on imaging. Definitive diagnosis is established when the mass is freely mobile within the IVC and right-sided cardiac chambers, without attachment to the endocardium or vascular wall.[11]

Prompt management is essential, as IVL can progress abruptly from an asymptomatic state to a life-threatening condition. Surgical excision remains the treatment of choice, aiming for complete tumor removal to minimize recurrence risk.[8] The surgical strategy—whether single-stage or two-stage—depends on the disease stage and the patient's overall condition. Tumors with IVC thrombus necessitate a collaborative effort between gynecologic and vascular surgeons, involving IVC incision without the need for cardiopulmonary bypass (CPB). Thrombus extending into heart chambers requires cardiothoracic surgical expertise, with CPB employed to minimize hemorrhagic risk during tumor extraction. While both single- and two-stage procedures are feasible, single-stage surgery generally offers superior outcomes, reducing the risk of tumor embolization between operations and lowering recurrence rates.[7]

Recurrence remains a significant clinical concern, as it may occur even decades after primary resection.[12] Some authors support adjuvant hormonal therapy postoperatively—using gonadotropin-releasing hormone (GnRH) analogs, aromatase inhibitors, or selective progesterone receptor modulators (SPRMs)—although evidence regarding its efficacy remains inconclusive.[12,13] Risk factors for recurrence include premenopausal status, incomplete tumor resection, and large initial tumor size, and hormonal therapy may be considered in such cases.[10] Preoperative administration of GnRH analogs can also be beneficial in reducing tumor size and facilitating resection in hormone-responsive IVL.[14] Postoperative surveillance lacks a standardized protocol, but most studies recommend follow-up imaging with CT at 3, 6, and 12 months post-surgery, followed by annual assessments to detect recurrence.[15] Rigorous and long-term follow-up remains essential due to the persistent risk of late recurrence.

CONCLUSION

The coexistence of intravascular leiomyomatosis (IVL) with smooth muscle tumour of uncertain malignant potential (STUMP) and intracardiac extension is exceedingly rare and poses significant diagnostic and therapeutic challenges. This case underscores the importance of maintaining a high index of suspicion in women presenting with large uterine masses and venous thrombus extending into the inferior vena cava or right atrium. Comprehensive preoperative imaging is crucial to delineate the full extent of disease and to facilitate meticulous surgical planning. Complete surgical excision through a multidisciplinary approach remains the cornerstone of management to prevent recurrence and life-threatening complications. Given the uncertain biological behavior of STUMP and the known risk of late recurrence in IVL, long-term postoperative surveillance is strongly recommended to ensure favorable outcomes.

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