Case Report

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A case report: intravenous leiomyomatosis extending from the uterus to the right atrium

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Introduction and importance: Intra veinous leiomyomatosis (IVL) is one pathology of a known group of ectopic leiomyomatosis that gathers many entities all defined by the presence of benign tumours arising from uterine smooth muscle cells. The authors aim through this case report to address the underdiagnosis of IVL in pre-menopausal women and the potential confusion with other cardiac tumours.

Case presentation: A 48-year-old woman initially treated for a suspected intracardiac myxoma underwent two surgeries. Seeking gynaecological care for menometrorrhagia related to a polymyomatous uterus, she was scheduled for radical surgery. Unexpectedly, extended explorations during a hysterectomy revealed a tumour originating from the pelvis, extending to the right atrium and inferior vena cava, indicative of intravascular leiomyomatosis.

Clinical discussion: IVL's diagnosis is often incidental during hysterectomy analysis, with symptoms mimicking uterine fibroids. Treatment involves radical surgery, emphasizing the importance of complete resection to reduce the significant risk of recurrence.

Conclusion: Pre-menopausal women with a history of hysterectomy or myomectomy and a detected right chamber mass should be screened for intravascular leiomyomatosis. Diagnosis relies on histological examination, guiding tailored treatment choices such as surgical resection with a focus on bilateral adnexectomy for optimal outcomes.

Keywords: case report, inferior vena cava, leiomyomatosis, myomatous uterus, myxoma

Introduction

Intravascular leiomymatosis known as IVL is a rare tumoral condition found in pre-menopausal women; it usually starts from a myomatous uterus and extends through the inferior vena cava to the cardiac tissue, the right atrium mostly, which can be confused as a myxoma.

Unlike IVL, Myxoma, which is known to be the most frequent cardiac tumour in adults is, in 80% of cases, located in the left atrium, rarely in the right atrium[1,2] and mostly discovered incidentally on echocardiography, as the clinical symptomatology is not specific.

Being a very rare pathology, only 300 cases of IVL have been described in the literature[3–6]. It is also poorly recognized in our medical environment and is often underdiagnosed, due to the tendency of our medical staff to focus on one single organ damage without further explorations. Whereas most same symptoms in the same area can be the translation of an extended disease.

Our purpose is to raise the importance of the diagnosis and treatment of this curable pathology in our medical community

HIGHLIGHTS

- A 48-year-old woman initially treated for suspected right atrial myxoma underwent two surgeries. Later diagnosed with intravenous leiomyomatosis (IVL), extending from the uterus to the right atrium.
- The patient presented with menometrorrhagia and pelvic heaviness, initially attributed to a polymyomatous uterus. The true extent of the pathology became evident during extended explorations after a disturbing event during pre-anaesthesia consultation for hysterectomy.
- IVL was challenging to diagnose, requiring transthoracic ultrasounds and thoracic abdominal pelvic computed tomography scans. The diagnostic certainty relied on histopathological analysis.
- The case necessitated a multidisciplinary approach involving intensive care, anaesthesiology, vascular surgeons, and gynaecologists due to the involvement of both the uterus and cardiac tissue.
- Treatment involved radical surgery, including total hysterectomy with bilateral adnexectomy. The risk of recurrence was highlighted, emphasizing the importance of complete resection.

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but to also add another case of intravenous leiomyomatosis of uterine origin with cardiac extension in literature, avoiding in the future, similar undiagnosed cases of women that are being dragged on for years without any real answers to their condition.

This work has been reported in line with the SCARE criteria[7].

Case presentation

The case we are sharing is about a 48-year-old woman mid-class stay-at-home, mother of three with one history of a miscarriage, a controlled high blood pressure using Calcium inhibitors and a balanced diet. The patient claims no food or drug allergies and no inherited disease.

The patient suffered from NYHA class III dyspnoea with explorations revealing what was initially suspected to be an atrial myxoma and had later undergone an intracardiac tumour resection on two occasions, both went under extracorporeal circulation.

An atrial myxoma was suspected due to the gelatinous aspect of the tumour.

Anatomopathological tests were made in an external laboratory on both occasions, and the patient was asked to, after complete recovery bring the laboratory results, but we had no feedback from her.

Five months later, the patient was received in the gynaecology department with pelvic heaviness and menometrorrhagia, pelvic ultrasounds showed a polymyomatous uterus, usually in these cases, our local gynaecology department does not necessarily ask for further exams in an effort to lessen the financial burden on the patients. (Table 1).

Clinical findings

In the pre-anaesthesia examination, the patient's check-up showed no abnormalities besides a distended abdomen, the patient was scheduled for a hysterectomy.

During the patient's monitoring, a rhythm disorder on the scope had been noticed including an inconstant P wave and irregular rhythm, blood pressure at 13/7 cmHg, and pulsed oxygen saturation at 99% without any sign of distress, which led our team to postpone the procedure for possible radiological exploration and re-evaluation of the cardiac function.

Diagnostic assessment and interpretation

The transthoracic ultrasounds performed showed a visible mass in the right atrium that reaches the inferior vena cava without any notable hypokinesia or valvopathy (Fig. 1).

An MRI objectified intra-pelvic leiomyomatosis (Fig. 2) while the thoracic abdominal pelvic computed tomography scan showed intracardiac extension passing by the inferior vena cava.

An abdominal tumoral origin with extraperitoneal extension reaching the inferior vena cava and the right atrium was then evoked.

Differential diagnoses

A completed thoracic, abdominal, and pelvic computed tomography (CT) scan reading was realized to exclude numerous differential diagnoses such as renal, retroperitoneal, or pelvic tumours in the first place but also other primary cardiac tumours such as myxoma or even thrombosis.

The myxoma diagnosis was doubtful since cardiac myxoma is usually in the left atrium, with no portion in the inferior vena cava.

Outcome and follow-up

To retain the IVL diagnosis we asked our patient to bring her previous anatomopathological where results showed that; immunostaining was positive for anti-smooth muscle actin antibodies (clone 1A4, DAKO).

anti H-caldesmon antibody (clone h-CD, DAKO), anti-desmin antibody (clone D33, DAKO).

The anatomopathological study showed an immuno-histochemical aspect confirming the histological hypothesis of smooth muscle differentiation.

In the absence of a clear malignant character at the histological stage, retaining two hypotheses: a disseminated intravascular leiomyomatosis or a benign metastasizing leiomyoma.

The surgical decision was postponed, and our patient's case was scheduled for a multidisciplinary staff including intensive care and anaesthesiology physicians, vascular surgeons, and gynaecologists.

During the multidisciplinary staff we came out with mainly two courses of action, either hysterectomy with bilateral adnexectomy or a hysterectomy with hormonal therapy, both followed by heart surgery. Due to low availability and the high cost of hormonal therapy we decided on going through with the former course of action.

The hysterectomy took place at Ibn Sina teaching hospital a week later after therapeutic management and the respect of the preoperative fasting rule after the implementation of a curative dose of enoxaparin-based anticoagulant treatment.

The patient had undergone her third open heart surgery a year later and was taken to the ICU for post-surgery car. The patient received venous thromboembolism prophylaxis after both surgery and didn't suffer from any complication or adverse event

Table 1

Timeline showing the sequence and order of events in the patient's history and presentation.

Timeline	Event
2020	First intracardiac surgery of a suspected right atrium Myxoma
2022	Second surgery of the recurrent right atrium Myxoma
Early 2023	The patient is showing signs of pelvic heaviness and menometrorrhagia, pelvic ultrasounds showed a polymyomatous uterus.
One week later	The patient is scheduled for a hysterectomy
Surgery due date	Surgery postponed due to a non-pre-existing cardiac rhythm disorder with no other alarming signs
20 days later	Histopathology final analysis confirmed intravascular leiomyomatosis, Hysterectomy surgery in the first place, patient was then scheduled for the heart surgery.

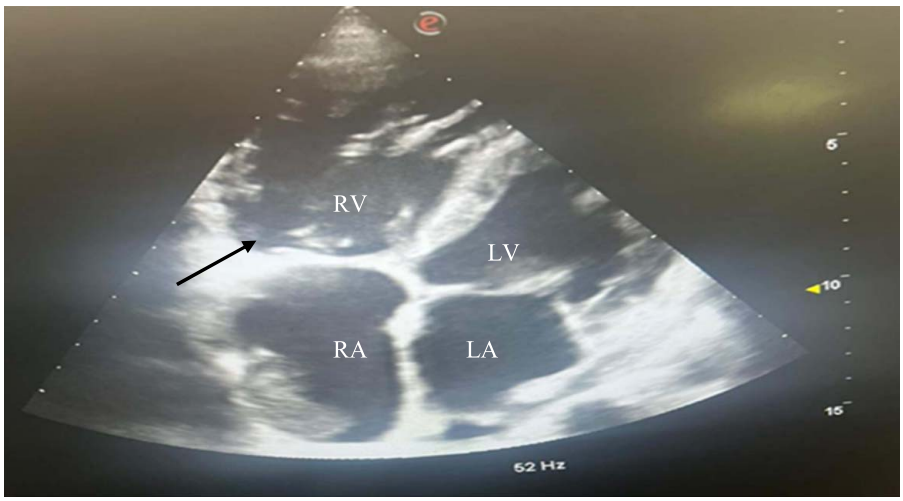

Figure 1. Intracardiac extension of intravenous leiomyomatosis. Transthoracic cardiac ultrasound: Apical 4-chamber section showing an elongated mass (black arrow) occupying the right atrium. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

while hospitalized. The patient was instructed during discharge to come for a follow-up 3 months later but we had no feedback from her.

Discussion

The search terms used were "Intravenous leiomyomatosis", "myxoma", "inferior vena cava", myomatous uterus", with the articles cited in this report being sourced mainly from the PubMed and ScienceDirect databases.

IVL was first described by Hirschfield in 1896[8].

It is a rare condition where only 200–300 cases have been reported in the literature[3–6].

Histologically, it consists of benign smooth muscle tumour cells growing within the uterine venous system[3–5].

IVL preferentially affects pre-menopausal women between 40 and 50 years old[3,5,6].

Two recent Asian studies (Chinese and Taiwanese) found IVL in 0.25[6] to 0.40%[3] of patients with uterine fibroids. This high prevalence may suggest that an ethnic risk factor may be involved

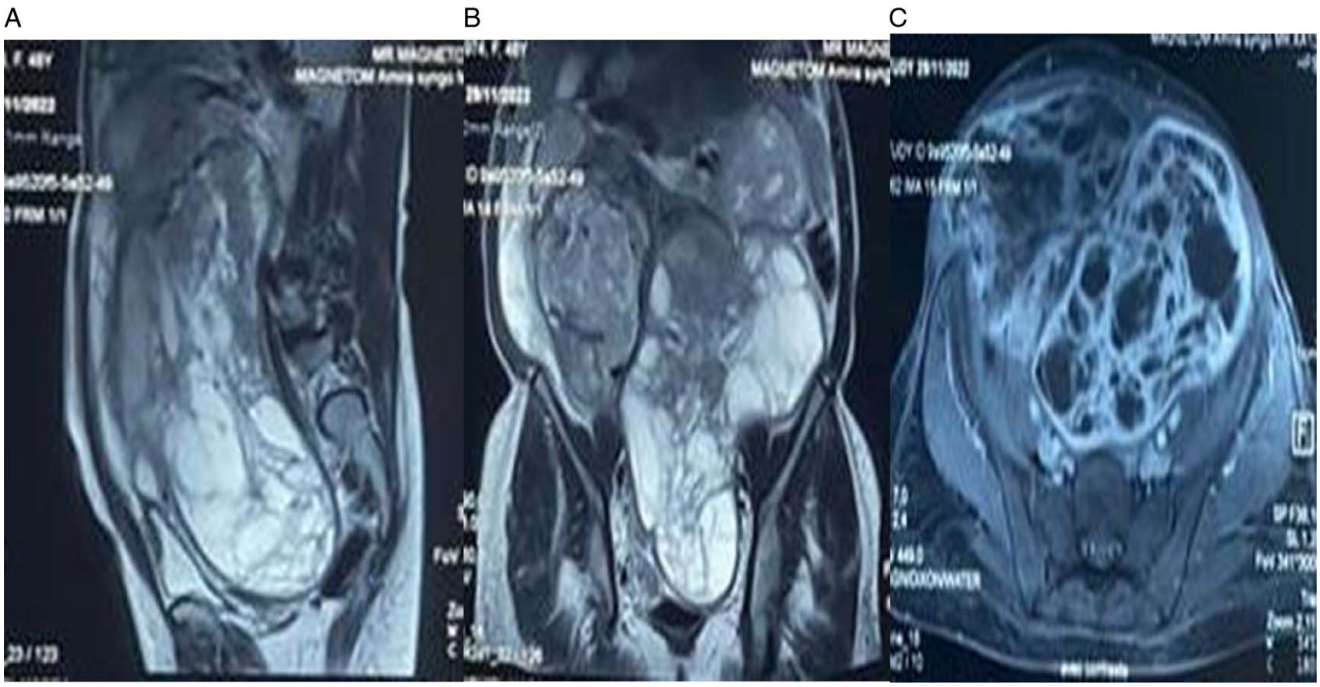

Figure 2. Uterine leiomyomatosis in cystic degeneration occupying the abdomino-pelvic cavity. coronal MRI T2 (A), sagittal T2 (B) and axial sequence after injection of gadolinium (C) Shows large masses with uterine origin, occupying the abdominal cavity, with heterogeneous signal, containing cystic areas in T1 hyposignal and T2 hypersignal, and tissue portions in diffusion hyposignal, homogeneously enhancing in relation to the myometrium.

in the development of the disease. However, there is most probably a recruitment bias since these publications come from reference centres for the disease.

The initial symptomatology of IVL, when it is located in the pelvis, is similar to that of uterine fibroids, mainly menometrorrhagia and pelvic heaviness with IVL symptoms becoming more pronounced and distinct as the condition progresses, especially if the tumour extends into the venous system, thus highlighting the importance of a thorough clinical evaluation or, in cases similar to ours, a thorough review of the patients medical and surgical history[6].

The diagnosis is then often made incidentally during the analysis of the hysterectomy specimen.

It consists of an invasion of the extra pelvic vessels (particularly the vena cava), affecting the venous return, which will be manifested in signs of right congestive heart failure: chest pain, dyspnoea, oedema of the lower limbs, syncope, and pulmonary embolism[4,5].

In case of cardiac damage along with obstruction of the tricuspid orifice, the risk is cardiac arrest and sudden death[2,5,9].

Paraclinical exams are often performed in front of noisy symptomatology (right heart failure in particular), confirming the diagnosis of IVL at an advanced stage[4,6].

Cardiac echocardiography allows the diagnosis of IVL if the cardiac extension is present.

In the context of women of childbearing age with a previous history or a synchronous uterine fibroid presenting a right intra-atrial mass originating from the vena cava, and without any argument for endothelial or endocardial invasion (mobile mass in the IVC and right atrium), the diagnosis of IVL must be suspected[9].

The tumour takes on the appearance of a loose, mobile serpentine mass in the vessels and in the heart[5,6,9], without invading the endothelium or the endocardium.

Certainty in diagnosis relies on histopathological analysis.

The treatment is based on radical surgery consisting of total hysterectomy with bilateral adnexectomy combined with complete resection of intravenous and intracardiac tumours[3,5,9].

Indeed, the risk of recurrence is directly correlated with the completeness of the resection[6,10].

In Ma's study of 76 cases, all 4 cases of recurrence occurred in stage 1 patients who refused hysterectomy with adnexectomy[10]. Thus, bilateral adnexectomy is recommended regardless of the stage of the disease.

The main risk of IVL is recurrence. According to the literature, the recurrence rate is 22.2[3] to 30%[2,5,6]. In the case of complete resection, it decreases to 7.6%[10].

We must also underline the weaknesses of our case report such as, the lack of long-term follow-up information and the limited discussion on the patient's perspective which would give this case report a more holistic view. We must also emphasize the biases in this case report, those mainly being a selection bias, as this study involves a single patient and the findings may not be representative of the broader population, and a generalization bias.

Conclusion

Intravascular leiomyomatosis screening should become a routine when, in pre-menopausal women, a history of hysterectomy or myomectomy is occurring along with right chamber mass showing in the imaging.

The definitive diagnosis relies on histological examination. This can be obtained by biopsies or by histological analysis of the surgical specimen. The treatment will be adapted according to age, location, size, comorbidities, and symptomatology. It is based on surgical resection and surgical castration (bilateral adnexectomy) or chemical castration (hormone therapy), with the former remaining the best attitude that comes with the best curative results and prognosis.

Ethical approval

This is a case report talking about a rare situation but derived from "standard" clinical practice so an ethics board approval was not required.

Consent

Written informed consent was obtained from the patient's next of kin to publish this report in accordance with the journal's patient consent policy.

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Author contribution

Writing the paper: Z.H. Data collection: N.E.I.J., Y.L. Proofreading: Z.S. Internal review: R.T. Contributors: S.L., N.O.

Conflicts of interest disclosure

The authors declare no competing interest.

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