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Successful multiple-step management of intravenous leiomyomatosis diagnosed after episode of acute abdominal pain: Case report and review of literature

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ABSTRACT

INTRODUCTION: We present the case of a 37-year old woman diagnosed with intravenous leiomyomatosis (IVL) that was managed uneventfully with multiple-step management.
PRESENTATION OF CASE: A 37-year-old woman was admitted because of acute abdominal pain. Emergency Computed Tomography demonstrated a big pelvic mass 5 × 15 cm of heterogenous composition intaking the contrast agent. Total hysterectomy with salpingoophorectomy was proposed to patient, however, patient expressed her will for fertility preservation and gave consent only for the resection of a single ovary. Laparotomy revealed the presence of myoma, multiple lesions of potential adenomyosis and cordon-shaped formations arising from uterus and extending mainly to left ovary. Final histological diagnosis was intravenous leiomyomatosis (IVL). MRI angiography revealed the presence of residual lesions in inferior vena cava. Laparoscopic resection was performed one month after laparotomy and left ovary was resected without complications. Venovenous bypass was finally performed three months later from initial surgery. The process was significantly labored, resulted in the successful resection of intravenous lesions but was complicated intraoperatively by right kidney rupture. After a follow-up of 33 months, case remains uncomplicated without signs or symptoms of potential recurrence.
DISCUSSION: Intravenous leiomyomatosis represents a rare clinical entity histologically bening but clinically aggressive. No consensus exists regarding the optimal management, especially in cases with initial will for fertility preservation.
CONCLUSION: IVL represents a rare clinical entity often presenting difficulties in diagnosis and optimal treatment. Large case-series studies should be encouraged to assess the optimal management.

1. Introduction

Intravenous leiomyomatosis (IVL) represents a rare clinical entity which could be defined as the extrauterine extension of smooth-muscle cells from a primary source, mainly intrauterine myoma, to the venous system. Indeed, IVL may affect anatomically neighbor veins such as intrauterine venules to much distanced anatomical structures, such as the iliac vein, inferior vena cava and even right heart chamber and pulmonary arteries [1,2]. The exact aitopathogenesis of IVL has not yet been exactly assessed; two main theories, however, have been: either the tumor arises from the walls of venous structures or the primary tumor (e.g., myoma) directly spreads into adjacent small venouses [3].

IVL has been characterized as a histologically benign entity with aggressive biological and clinical characters. On the one side, IVL consists of smooth muscle tissue with no malignancy histopathological characters [4]. However, extrauterine extension of cells may lead to invasion of organs such as heart, liver which may result in, dyspnea, cardiac murmur, Budd-chiari syndrome, syncope episodes and even death [5,6]. Therefore, it is essential to achieve the complete resection of IVL as the potential of recurrence may be apparent. However, search of the Pubmed database by using the term "intravenous leiomyomatosis" in title or abstract demonstrates exactly 276 published case reports or cases series, therefore, indicating the rarity of the disease. As a result, no consensus has yet been achieved regarding the optimal diagnostic approach or either management of IVL.

We present the case of a 37-year-old woman that was diagnosed with intravenous leiomyomatosis and was finally managed with a multiple-step procedure, her follow-up being heretofore uncomplicated.

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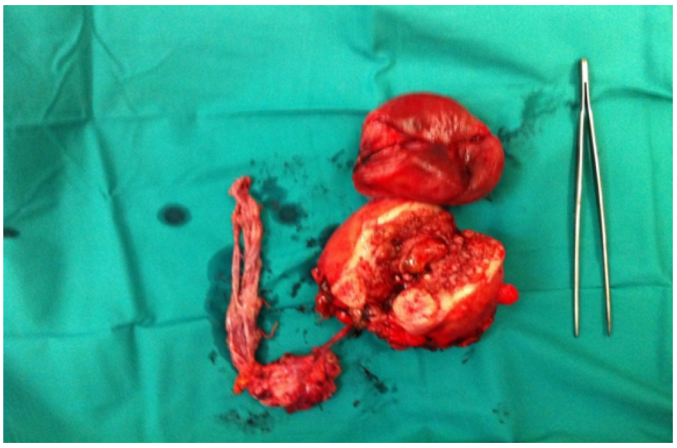

Image 1. Cordon-shaped formations arising from uterus and being extended mainly to left ovary.

2. Case report

A 37-year-old woman was admitted to Emergency Unit of Interbalkan Medical Center of Thessaloniki because of acute abdominal pain. The patient reported initiation of her symptoms one week earlier, the symptoms becoming very intense during the previous four hours. The patient's medical history revealed no remarkable pathology. The woman had already delivered two uncomplicated pregnancies with natural deliveries. The patient only reported from her gynaecological history the presence of a submucosal myoma that was diagnosed ten years earlier and which was followed-up with ultrasound on an annual basis, being stable in size and without remarkable symptoms apart from rare episodes of mild pelvic pain.

Clinical examination of the patient revealed signs of peritoneal inflammation with positive rebound testing. Laboratory examination indicated elevated white blood cell count of about 16,000/mL of which neutrophile type was 80.1%, while CRP value was also increased (2.1 gr/dL). Ultrasound examination confirmed the presence of a huge subcutaneous myoma (over 10 cm size) and reactive liquid in Douglas and Morrison anatomical space, without however, providing further imaging information about the exact cause of abdominal pain. Emergency Computed Tomography was performed, demonstrating a large pelvic mass with dimensions 5×15 cm with heterogenous composition, intaking the contrast agent. The mass arose from the uterus but was also extended to both ovaries, mainly in the right one. There was also presence of liquid in both Douglas and Morrison anatomical spaces, while CT also reported retroperitoneal lymph nodes of about 1 cm. The final CT diagnosis was myomatous uterous with high suspicion of malignancy.

The decision of total hysterectomy with salpingoophorectomy was made. The patient, however, expressed her desire for fertility preservation and gave consent to proceed only to the resection of one ovary unless the diagnosis of endometrial cancer was intraoperatively made by frozen section biopsy. Laparotomy was performed that revealed the presence of myoma, multiple lesions of potential adenomyosis and cordon-shaped formations arising from the uterus and extending mainly to the left ovary. Frozen section biopsy was negative for potential malignancy and the surgery was completed with the resection of uterus and left ovary (Image 1). Final histological diagnosis was intravenous leiomyomatosis (IVL). The patient was explained that IVL does not represent a histologically malignant entity, but as disease is associated with high risk of recurrence and aggressive clinical behavior, the patient was advised to resect the right ovary as well. Furthermore, the patient was advised to perform MRI angiography, which revealed the presence of residual lesions in inferior vena cava. Therefore, the therapeutic plan that was finally decided was to perform laparoscopic resection of left ovary and, with the recommendation of the vascular surgeon, to perform a venovenous bypass. Laparoscopic resection was performed one month after laparotomy and left ovary was resected without complications. A venovenous bypass was finally performed three months from the initial surgery. The process was significantly labored, resulting in the successful resection of intravenous lesions but was complicated intraoperatively by rupture of the right kidney. The kidney rupture was managed with pig-tail placement for a 6-month interval, after which kidney function was totally restored.

The patient was afterwards set in follow-up with clinical, laboratory and MRI imaging every 6 months for the first two years and on an annual basis afterwards. After an interval of 33 months, the case remains uncomplicated without signs or symptoms of potential recurrence.

3. Discussion

We presented the case of a 37-year-old woman that was diagnosed with intravenous leiomyomatosis and was successfully treated with a multiple-step surgical approach.

IVL represents a histologically benign entity with malignant clinical behavior. Indeed, there is no reported case of IVL finally progressing to malignancy. However, IVL may cause remarkable systematic complications, presents significant diagnostic difficulties and is also characterized by a relatively increased possibility of recurrence. Indeed, the expansion of IVL through venous vessels may affect even the heart or liver, thereby leading in severe complications such as syncopal episodes, pulmonary embolism, and sudden death [7–9]. Furthermore, the reported recurrence rates range between 15 and 30%, while cases of recurrence have been mentioned from 6 months to even 15 years later [4,10]. Given the fact that no consensus has also been made about the optimal follow-up strategy, IVL may pose significant challenges in an effort to prevent recurrence of the disease.

Diagnosis of IVL is also an issue of high clinical difficulty. IVL should be differentially diagnosed from leiomyosarcoma, which represents a malignant clinical entity. There have been reports of leiomyosarcomas presented with similar signs and symptoms with that of IVL [11–13]. The issue of diagnosis may also be more complicated by the fact that no imaging method has been reported to have high specificity for IVL. This was also the case regarding our patient; the combination of ultrasound and computed tomography posed a high suspicion of uterine malignancy rather than the diagnosis of IVL. Furthermore, histological diagnosis based on frozen section intraoperatively may also not be determinant. Therefore, final diagnosis should be based only on the postoperative examination of the whole surgical tissue [5]; even this may result in multiple surgical procedures especially as IVL is presented in women with a desire for fertility preservation. Moreover, especially in cases of intracardiac expansion, differential diagnosis of IVL should also include thrombus-in-transit, which may also appear as mobile masses of venous casts, giving a "popcorn" appearance to the cardiac chambers [14,15].

The crucial point about IVL is the optimal therapeutic approach. There are two separate approaches to this issue. The first option is the radical surgical approach. The radical surgical procedure seems to be curative either in one or in multiple steps [5]. Hysterectomy and bilateral salpingoophorectomy have been proposed as the gold standard method of treatment by several studies. There are also cases in which radical instead of total hysterectomy should be considered [16]. Apart from gynaecological surgery, vascular sur-

gical procedures that have been proposed in an effort to radically resect disease may enroll venovenous bypass and replacement of affected vessel with appropriate graft. As the review of literature indicates, a relatively restricted number of published cases with often heterogenous clinical manifestations and complications, and no randomized controlled trials have been performed, in order to assess whether surgery should be performed in one or in multiple steps. It is a fact that coexisting complications such as coagulopathy or severe haemorrage should rather discourage the potential of the one-step procedure [5]. In our case, the desire of patient to maintain fertility and the fact that the disease did not macroscopically seem to expand to large vessels, which was finally diagnosed only with MRI imaging, led to a multiple-stage procedure. However, in case the diagnosis is apparently made without patient presenting acute symptoms or complications, the one-step procedure could be recommended.

Apart from surgical resection, however, usage of selective estrogen receptor modulators has been recently proposed as an alternative approach to treat IVL. The effect of raloxifaine on reducing the size of uterine leiomyoma has been indicated, but effectiveness against IVL has only been examined in animal models [3,16]. Furthermore, antioestrogens such as tamoxifaine have been implicated in the treatment of cases with residual disease with controversial results [3,17]. Therefore, it seems that surgical resection of IVL either in one-step or in multiple-step strategy remains the optimal therapeutic approach regarding a clinical entity with few reported cases in which research about optimal diagnosis and treatment still leaves a lot to be desired.

In conclusion, IVL represents a rare clinical entity often presenting difficulties in diagnosis and optimal treatment. Large multi-center case studies should be encouraged in order to improve our level of knowledge and thereafter quality of provided health services for a disease which may be characterized as histologically benign but clinically aggressive.

Conflict of interest

None.

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Ethical approval

None.

Consent

Informed and written consent has been given by patient.

Authors contribution

C.E. conceived the basic concept and reviewed the manuscript. S.P, C.M-S and I.K wrote the inital draft. M.G., A.I. and I.T reviewed the manuscript.

Guarantor

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