

Case Report

Intravascular Leiomyomatosis: A Case Report and Review of the Literature

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Abstract

Intravenous Leiomyomatosis (IVL) is a rare tumor which affects, most frequently, middle aged premenopausal women with previous history of uterine leiomyoma. We report the case of a 46-year old woman with an intravenous leiomyomatosis growing within the Left Gonadal Vein (LGV), Left Renal Vein (LRV) and suprarenal Inferior Vena Cava (IVC).

Keywords: Intravenous leiomyomatosis; Left gonadal vein; Left renal vein

Introduction

Intravenous Leiomyomatosis (IVL) is a rare tumor and a rare variant of a uterine leiomyoma which slowly grows invading the extra uterine venous system and into the Inferior Vena Cava (IVC), right atrium and pulmonary arteries [1-14]. It was first described by Hirschfield in 1896 and defined by Norris and Parmly in 1975 [2] affecting a wide range of ages, between 21 and 86 years of age [4]. It is normally diagnosed incidentally in patients with history of leiomyoma and it can occur within a leiomyoma or in the absence of it. It is characterized by a growth of benign mature smooth muscle cells in the lumen of veins and lymphatic vessels. It is a benign entity, but because of its behavior, it could be considered malignant, since cases of intracaveal and intracardiac extension, and even lung metastases, have been described. Extension, in some cases, into the cardiopulmonary system causing dyspnoea, syncope, congestive heart failure or sudden death may occur. Its treatment with complete surgical resection is important due to its aggressive behavior and extension [1-16]. We report the second case of intravenous leiomyomatosis treated in our Vascular Surgery Department.

Case Report

The patient is a 46-year old woman with a previous history of leiomyoma who needed hysterectomy. In the histopathological result, intravascular leiomyomatosis was diagnosed. In preoperative Computerized Tomography (CT) scan, there is a finding of a mass occupying the Inferior Vena Cava (IVC) which causes doubts whether it could be a thrombus or an intravascular leiomyomatosis.

After serial CT scans, Magnetic Resonance Imaging (MRI) and Positron Emission Tomography (PET) scan, diagnosis of an intravenous growing mass extending from Left Gonadal Vein (LGV), into the Left Renal Vein (LRV) and suprarenal Inferior Vena Cava (IVC) in venous-phase Computerized Tomography (CT) scan during her follow-up, was performed. In a multidisciplinary session with Gynaecologists, total surgical resection was decided.

Surgery was performed by our team of Vascular Surgeons in collaboration with Urology. Under general anaesthesia and epidural catheter, bilateral subcostal laparotomy was performed, with control of LVG, LRV, suprarenal IVC and left renal artery. Right ovary was found in the origin of the tumor in pelvis, with drainage into LGV (Figure 1). Implantation of the tumor was

also observed inside a lumbar vein. Proximal and distal control of the tumor was performed. Venotomy of LRV at the drainage of LGV. Resection of tumor was gently carried out, with ligation of lumbar vein and LGV. Care was taken by controlling IVC to prevent embolization during extraction of the tumor (Figures 2 and 3). Anexectomy of right ovary was also performed.

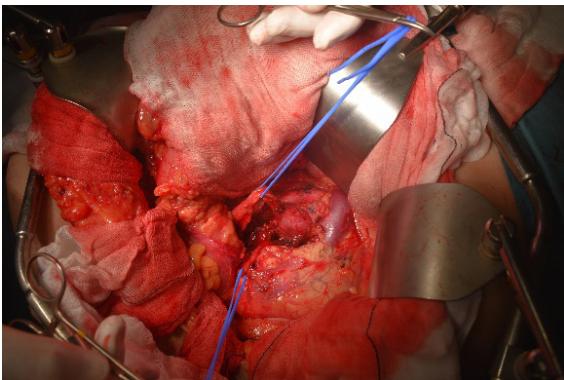


Figure 1: Right ovary in the origin of the tumor in pelvis, with drainage into LGV, before performing anexectomy.

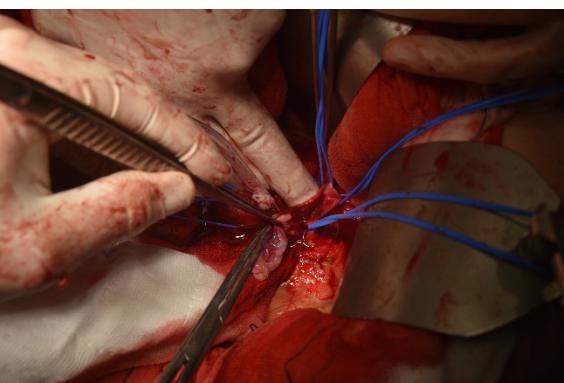


Figure 2: Venotomy of LRV at the drainage of LGV and careful resection of tumor which was not adhered to the venous wall.

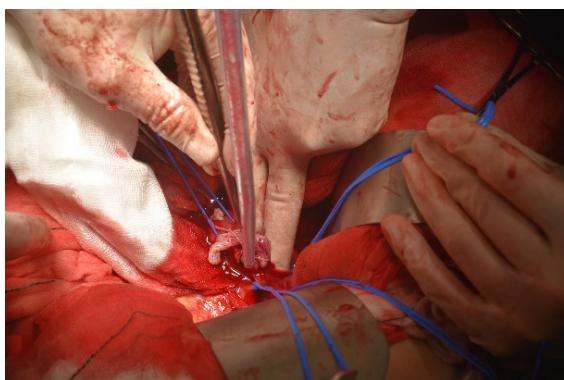


Figure 3: Venotomy of LRV at the drainage of LGV and careful resection of tumor which was not adhered to the venous wall.

The piece of tumor of 13 cm of length (Figure 4) and the right ovary were sent for histopathological examination, with the result of an IVL without abnormal mitotic activity and ovary without significant findings.



Figure 4: Tumor, LGV, lumbar vein and ovary resected.

The patient was discharged after 10 days, with no residual tumor in CT scan performed three months after discharge.

Discussion

IVL is a rare condition which affects, most frequently, middle aged premenopausal women between 30-50 years with previous history of uterine leiomyoma and that are multiparous [8].

Less than 300 cases have been reported in literature [11]. It is histologically benign, composed by smooth muscle cells and has expression of hormonal receptors with clear response to hormone influence, just as it happens with leiomyomas. There are two theories which try to explain the pathogenesis of IVL: one supports an origin from the venous wall, and, another, as an extension of a uterine leiomyoma into the uterine venule. Most studies support its origin from a uterine leiomyoma given its coexistence [2,5,9]. Despite its histological characteristic, its growing pattern is aggressive, extending from pelvic veins into IVC and growing within venous channels into the right atrium and pulmonary arteries. This last entity is defined as intracardiac leiomyomatosis (10% of cases) and can have fatal consequences [11]. A case of intravenous leiomyomatosis with cardiac extension was treated at out department and reported in literature, with a successful resection of the tumor performed in one stage through midline sternotomy and midline laparotomy [13].

Because it is so uncommon, it can be misdiagnosed due to the variety of symptoms and can be mistaken with other more frequent entities such as venous thrombosis and atrial mixoma if there is cardiac extension. The most common symptoms are related to mass effect and venous obstruction, such as abdominal or pelvic pain, lower extremity swelling and more rarely with

thromboembolic events [16]. A correct differenced diagnosis must be performed with other tumor which invade IVC and LRV such as renal tumours and leiomyosarcoma of Vena Cava [2,5,13]. Tumor markers should be analysed.

It is important to perform an extension study, with Magnetic Resonance Imaging (MRI), venous-phase CT scan, transthoracic and trans oesophageal echocardiography. This will help us to plan surgery in a multidisciplinary way [13].

Ma, et al. described the staging system for this tumor and divides it into 4 stages.

Stage 1: the tumor penetrates the uterine venous wall but is confined in the pelvic cavity.

Stage 2: the tumor extends into the abdominal cavity but does not reach the LRV.

Stage 3: the tumor reaches the LRV and IVC and extends into the right atrium but does not reach the pulmonary artery.

Stage 4: the tumor reaches the pulmonary artery or there are lung metastases [7].

A primary approach with hysterectomy with double salpingoophorectomy must be performed [5,8]. Complete surgical resection is up to date the gold standard. Its early treatment is important in order to prevent intracardiac invasion with subsequent fatal consequences [3,5,7-16].

Recurrence rate is reported to be approximately 30% due to incomplete resection. Some authors support the additional treatment with hormone therapy using gonadotropin-releasing hormone agonists in patients with incomplete tumor resection, to prevent recurrence, although this has not been yet demonstrated [3,5-8].

Conclusion

IVL is a rare histological benign tumor with an aggressive pattern which can infiltrate venous vessels and extend into cardiac chambers causing fatal consequences. Its correct diagnosis is important along with an early surgical complete removal. A correct differenced diagnosis must be performed with other tumor which invade IVC and LRV such as renal tumours and leiomyosarcoma of Vena Cava. It should be considered in patients presenting with uterine leiomyoma and venous thrombosis as a possible diagnosis due to its behavior with venous occupation. In cases where incomplete resection is performed, there could be a place for hormone therapy in order to avoid recurrences, although this has not yet been demonstrated.

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