International Journal of Academic Health and Medical Research (IJAHMR)
ISSN: 2643-9824
Vol. 8 Issue 2 February - 2024, Pages: 179-184

Intravenous leiomyomatosis, one of the extra uterine forms of leiomyoma: report of a case and review of the literature:

YACOUBI KHEBIZA S[1], Jayi S[2],TAHIRI L[3] , MOUHCINE Y[4] , BOUBOU M[5] , FZ FDILI[6], Chaara H[7], Melhouf MY A[8]

Department of Obstetrics Gynecology II CHU Hassan II of Fez. (1,2,6,7,8)
Anatomopathology department, CHU HASSAN II of Fez.(3)
Radiology department of mother and child hospital , CHU HASSAN II of Fez .(4,5)

Abstract: Intravenous leiomyomatosis is one of unusual uterus benign tumor . In this article, we report a case of intravenous leiomyomatosis discovered postoperatively in a patient initially operated on for suspicion of uterine sarcoma. through this work, we will expose the clinical and radiological signs, the grades of this pathology, as well as the management, the prognosis and the risk factors for recurrence in the event of this rare form of pathology being affected.

Keywords: Intravenous leiomyomatosis , surgery , recurrence .

INTRODUCTION

Uterine leiomyoma is the most frequently encountered benign tumor in women with an estimated frequency of 30% in patients aged over 35 years.

Extra-uterine forms (called leiomyomatosis) have been described.

Histologically, they correspond to a typical uterine leiomyoma with unusual locations (outside the genitourinary tract) [1,2].

Four distinct entities are reported: intravenous leiomyomatosis , benign metastatic leiomyoma , disseminated peritoneal leiomyomatosis ,hereditary leiomyomatosis .

Although benign, these forms of leiomyomatosis present aggressive characteristics, primarily suggesting a malignant pathology (leiomyosarcoma for disseminated peritoneal leiomyomatosis in particular). [3,4].

Only anatomo-pathological analysis makes it possible to confirm the diagnosis and therefore benignity.

Of unknown etiology, their evolution is indolent and hormone-dependent

These ectopic forms are often unrecognized or diagnosed late, leading to inappropriate or late treatment.

Through this presentation we will present a case of intravenous leiomyomatosis discovered on anapathic specimen after carrying out a hysterectomy for suspicion of uterine sarcoma followed by a review of the literature

1- CLINICAL CASE

• This is a 41-year-old female patient, nulliparous and still menstruating, with no particular personal pathological history.

Family history: a sister operated for hysterectomy at a young age whose anatomopathological study was benign according to the patient (no proof)

The patient consults for management of moderate chronic abdominal-pelvic pain, such as heaviness, associated with light abnormal uterine bleeding.

The physical examination finds a patient with OMS at 0 and BMI at 22, with on the gynecological level: on the speculum an aspirated cervix of macroscopically normal appearance with stigmata of bleeding and on Vaginal Touch coupled with abdominal palpation a cervix and vaginal walls soft, mass filling the posterior anterior and lateral CDS, uterus of size difficult to assess with mass reaching as far as the epigastrium difficult to mobilize in relation to the deep plane.

Paraclinical examination

Pelvic ultrasound :

Uterus increased in size reaching the xiphoid appendage; Myometrium is the site of several myomatous nuclei, most of which are heterogeneous without obvious vascularity, types 03-04 and 05

Endometrium followed fine with a 25 mm intracavitary corporeal image non-vascularized (polyp? myoma?), with presence of hematometry;

Both ovaries not seen

No effusion.

Thoraco abdomino pelvic scanner: figure 1

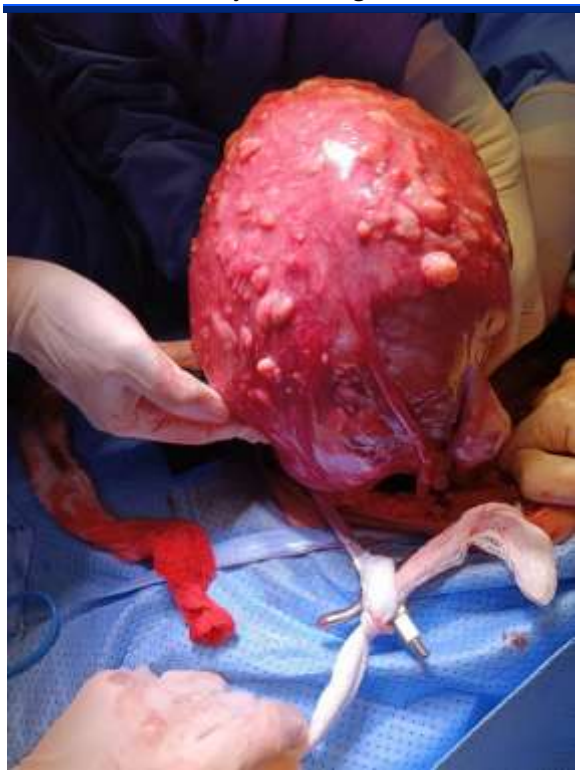
Enlarged uterus, polymyomatous with reworked myomas requiring comparison with MRI data to eliminate signs of atypia;
Bilateral external iliac adenomegaly.
A few pulmonary parenchymal nodules not exceeding 4.5 mm to be monitored given the context
Absence of suspicious ADP
Absence of pericardial or pleural effusion

Pelvic MRI:

Large polymyomatous uterus, with myomas remodeled into hyaline degeneration,
hemorrhagic, hypercellular and atypical necrobiosis
Pedunculated myometrial lesion, left lateral anterior isthmic, measuring 8.5 x 6.5 cm with a pedicle measuring 4 cm. This lesion is suspicious for malignancy, it presents nodular contours with a focal discontinuity of the serosa at its postero-inferior pole;
Bilateral external iliac atypical adenomegaly

figure 1 : **abdominopelvic scan in axial section; sagittal and coronal, showing: Uterus increased in size, measuring 30 x 12.5 cm on a sagittal section, seat of countless myometrial lesions, the majority are liquefied (yellow star), well limited, presenting peripheral enhancement (green arrow) of which the larger is anterior isthmic (red circle), less than 50% intra myometrial measuring 07 x 08 cm and which rests on the bladder, others are enhanced intensely and others have poorly limited contours.**

Figure 2 : intraoperative appearance of the uterus



- **Histological assessment:**

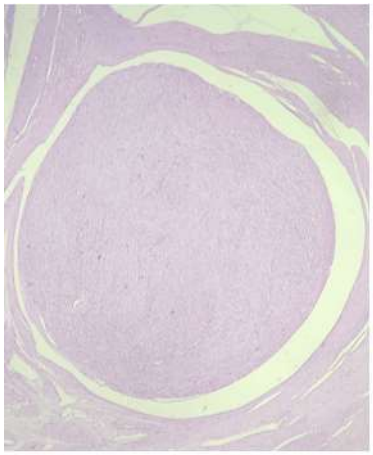
Performance of an inter-adnexal hysterectomy with bilateral salpingectomy and bilateral pelvic dissection Simple surgical results

Anapath results:

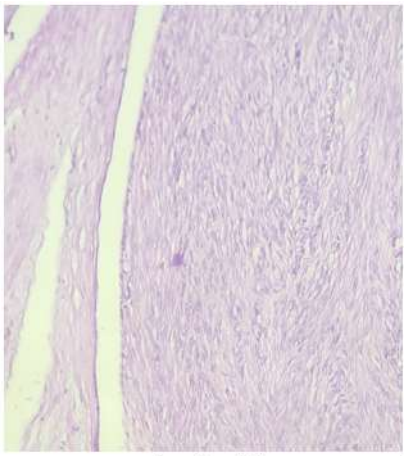
- Intravenous leiomyomatosis, cervix, endometrium and the 2 tubes are normal in appearance,
-right dissection: finds 5 leiomyomatous nodules with 6 lymph nodes in reactive lymphadenitis
-left dissection: 3 leiomyomatous nodules with 6 lymph nodes in reactive lymphadenitis

Figure 3 macroscopic appearance of the hysterectomy specimen

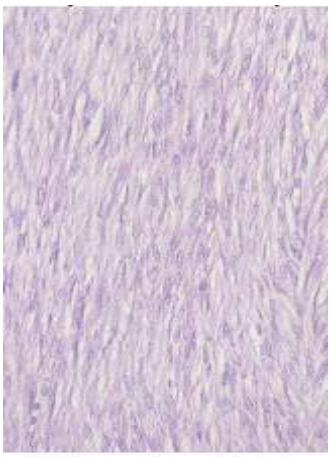
Figure 4 :



HESx200: intra venous well limited proliferation



HESx200: spindle cell proliferation arranged in interlacing bundles cell



HESx400: spindle tumor cells with no atypia, necrosis or mitotic figures

2- DISCUSSION

a- Epidemiology and etiology

Intravenous leiomyomatosis is a rare benign smooth muscle tumor proliferation, encountered exclusively in white women, with a history of hysterectomy for uterine fibromyomas.

IVL preferentially affects pre-menopausal women between 40 and 50 years old

The etiology of IVL remains uncertain but two hypotheses are mainly described: [3,4].

- by hematogenous dissemination with invasion of the vessels by a uterine leiomyoma
- by metaplasia (the tumor would come directly from the venous endothelium)

b- clinical signs :

IVL can spread into the venous circulation from two distinct venous routes:

1st route: the uterine vein:

its extension is along the internal and common iliac veins then into the IVC

2nd way the ovarian vein:

the extension will be done directly into the IVC on the right and via the left renal vein on the left, without affecting the iliac veins

Once the IVC is reached, the pathology can extend to the right atrium (intracardiac leiomyomatosis) and to the pulmonary arteries (cases have been described). There are also forms associated with pulmonary nodules

Cardiac waiting represents the extreme form of IVL [5,6].

The clinical signs depend on whether the localization is limited to the pelvis or whether it extends beyond:

- Limited to the pelvis: symptomatology similar to that of uterine fibroid, mainly meno-metrorrhagia and pelvic heaviness. The diagnosis is then often made incidentally during analysis of the hysterectomy specimen.

- invasion of the extra pelvic vessels (particularly vena cava) an abnormality of venous return, signs of right-sided congestive heart failure: chest pain, dyspnea, edema of the lower limbs, syncope, pulmonary embolism.

In the event of cardiac damage, with obstruction of the tricuspid orifice, the risk is cardiac arrest and sudden death.

c- **radiological diagnosis:**

thoraco abdomino pelvic scan, MRI: we can classify the tumor, complete with transthoracic ultrasound if cardiac damage is suspected,

There are 4 different grades of intravenous leiomyomatosis depending on the degree of invasion of the tumor.

- Grade 1: corresponds to invasion of the uterine vessels by the tumor without extra-pelvic invasion.
- Grade 2 is extension into the abdominal cavity without involvement of the renal vein.
- Grade 3 the renal vein and the inferior vena cava as well as the right atrium are invaded.
- grade 4 corresponds to damage to the pulmonary arteries and/or the presence of pulmonary metastases

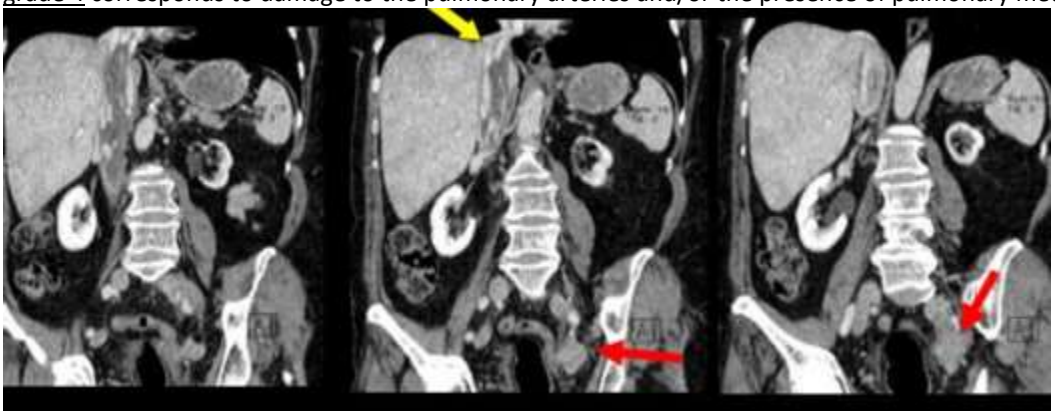


Figure 5 localization of leiomyomatosis in the inferior vena cava

d- Histology:

The definitive diagnosis is based on anatomopathological analysis!!!.
Pre-operative biopsies are not performed systematically, particularly in cases of typical clinical and ultrasound presentation.
The tumor is composed of smooth muscle cells normally found in the uterus.
Microscopically, the tumor is positive for actin and desmin and
we also note the presence of hormonal receptors for progesterone and estrogen.
Mitotic analysis reveals low activity[7,8].

e-differential diagnosis

- endovenous tumor buds of kidney cancer, Wilms tumor, adrenocorticaloma
- primary leiomyosarcoma of the inferior vena cava
- malignant non-Hodgkin lymphoma and soft tissue sarcoma
- retroperitoneal fibrosis

f- treatment

Ideally, treatment is based on radical surgery consisting of total hysterectomy with bilateral adnexectomy associated with complete resection of intravenous and intracardiac tumors.

The benefit of adnexectomy is explained by the hormone sensitivity of the tumor. Thus, surgical castration is indicated in order to reduce the risk of recurrence, particularly in the event of incomplete resection, especially in pre-menopausal patients.

In the event of an incidental diagnosis of IVL on the hysterectomy specimen, it is necessary to perform additional **surgical** (bilateral adnexectomy) or **chemical** (hormonotherapy) castration.
Chemical castaration with GnRH (Gonadotropin releasing hormone) analogues (a-GnRH), selective estrogen receptor modulators (SERMs) such as tamoxifen and anti-aromatases (AA).

In young patients, chemical castration with a-GnRH will be preferred to surgical castration.
In older patients, AA will be preferred in case of refusal or impossibility to perform a bilateral adnexectomy. [8,9].

g- follow up

The main risk of IVL is recurrence.

According to the literature, the recidivism rate is 22.2% to 30%, in case of complete resection it decreases to 7.6%.
The risk factors for recurrence are as follows: young age, the initial size of the tumor, an incomplete resection without postoperative hormonal therapy, uterine and ovarian preservation,
Recurrence can occur several years after surgery.

It may consist of a venous or abdominal recurrence with multiple pelvic masses.
In the event of recurrence, further radical surgery with complete resection significantly improves the prognosis. [9].

h- **monitoring:**

Long-term monitoring is recommended.
It is based on CT or magnetic resonance imaging (MRI) between 3 and 6 months after surgery then regularly every 2 to 5 years, depending on the grade and completeness of the resection.

So for our patient given the incomplete nature of the surgery:
We can offer our patient either a bilateral oophorectomy or chemical castration
Then install surveillance
our patient refused surgical revision, so we put her under chemical castration[9].

8- Conclusion:

Intravenous leiomyomatosis is a rare pathology.
It mainly affects pre-menopausal women with a history of uterine myoma works or not.
The definitive diagnosis is histological. Find out most of the time about hysterectomy piece.
Treatment is based on resection surgery and surgical (bilateral adnexectomy) or chemical (hormonotherapy) castration.
Follow-up must be closer and prolonged due to their high risk of recurrence.

Bibliography:

1. Fasih N, Prasad Shanbhogue AK, Macdonald DB, Fraser-Hill MA, Papadatos D, Kielar AZ, et al. Leiomyomas beyond the uterus: unusual locations, rare manifestations. Radiogr Rev Publ Radiol Soc N Am Inc. 2008;28: 1931–1948. doi:10.1148/rg.287085095
2. Low H-Y, Zhao Y, Huang K-S, Shen H-P, Wu P-J, Tseng C-J. Intravenous leiomyomatosis of the uterus: A clinicopathological analysis of nine cases and literature review. Taiwan J Obstet Gynecol. 2017;56: 362–365. doi:10.1016/j.tjog.2017.04.017
3. Mahmoud MS, Desai K, Nezhat FR. Leiomyomas beyond the uterus; benign metastasizing leiomyomatosis with paraaortic metastasizing endometriosis and intravenous leiomyomatosis: a case series and review of the literature. Arch Gynecol Obstet. 2015;291: 223–230. doi:10.1007/s00404-014-3356-8
4. Lin J, Song X, Liu C. Pelvic intravascular leiomyomatosis associated with benign pulmonary metastasizing leiomyoma: clinicopathologic, clonality, and copy number variance analysis. Int J Gynecol Pathol Off J Int Soc Gynecol Pathol. 2014;33: 140–145. doi:10.1097/PGP.0b013e31828def26
5. Valdés Devesa V, Conley CR, Stone WM, Collins JM, Magrina JF. Update on intravenous leiomyomatosis: report of five patients and literature review. Eur J Obstet Gynecol Reprod Biol. 2013;171: 209–213. doi:10.1016/j.ejogrb.2013.09.031
6. Zhang G, Yu X, Lang J. Intravenous leiomyomatosis with inferior vena cava or intracardiac extension and concurrent bilateral multiple pulmonary nodules: A report of 2 cases. Medicine (Baltimore). 2016;95: e4722. doi:10.1097/MD.0000000000004722
7. Li R, Shen Y, Sun Y, Zhang C, Yang Y, Yang J, et al. Intravenous leiomyomatosis with intracardiac extension: echocardiographic study and literature review. Tex Heart Inst J. 2014;41: 502–506. doi:10.14503/THIJ-13-3533
8. Ma G, Miao Q, Liu X, Zhang C, Liu J, Zheng Y, et al. Different surgical strategies of patients with intravenous leiomyomatosis. Medicine (Baltimore). 2016;95: e4902. doi:10.1097/MD.0000000000004902
9. Wolfe S, Sullivan J, Kahn L. Perivascular Leiomyomatosis: A Unique Case Report. Int J Surg Pathol. 2018; 1066896918772340. doi:10.1177/1066896918772340
10. Kim Y-N, Eoh KJ, Lee J-Y, Nam EJ, Kim S, Kim SW, et al. Aberrant uterine leiomyomas with extrauterine manifestation: intravenous leiomyomatosis and benign metastasizing leiomyomas. Obstet GynecoSci. 2018;61: 509–519. doi:10.5468/ogs.2018.61.4.509