Case Report

iMedPub Journals
http://www.imedpub.com/

Journal of Rare Disorders: Diagnosis & Therapy

2016
Vol.2 No.3:44

Intravenous Leiomyoma with Extension to Heart: A Case Report

Khan Mohammed Firoj[1], Shanmukha Sasank[2], Hai bin Yu[1] and Xian en Fa[1]

[1]Department of Cardio-vascular Surgery, 2nd Affiliated Hospital of Zhengzhou University, Zhengzhou, Henan, China

[2]Department of Cardio-vascular Surgery, 1st Affiliated Hospital of Zhengzhou University, Zhengzhou, Henan, China

Corresponding author: Khan Mohammed Firoj, Department of Cardio-vascular Surgery, 2nd Affiliated Hospital of Zhengzhou University, Zhengzhou, Henan, China, Tel: 86130277787275; E-mail: falishakhan72@yahoo.com

Received date: March 11, 2016; **Accepted date:** May 21, 2016; **Published date:** May 25, 2016

Citation: Firoj KM, Sasank S, bin Yu H, et al. Intravenous Leiomyoma with Extension to Heart: A Case Report. J Rare Dis Diagn Ther. 2016, 2:3.

Abstract

Leiomyoma is a benign smooth muscle tumor that very rarely becomes cancer. Uterine leiomyoma or uterine fibroid is a benign tumor of uterus arising from smooth muscle cells of myometrium. Intravenous leiomyoma with intracardiac extension is a very rare clinical entity usually develops exclusively in women arising from either uterine venous wall or uterine leiomyoma [1] which is commonly misdiagnosed as primary cardiac tumor, such as Myxoma. We report the case of 48 year old woman with intravenous leiomyoma arising from left common iliac vein and extended to the heart through the inferior vena cava presented with symptoms of hemodynamic disorder. The patient had the history of uterine fibroid. Thoraco-abdominal CT was performed to finalize the diagnosis. The intravenous tumor with its extension to the right atrium was ultimately resected via cardiotomy without the need of prolonged hypothermia during cardiopulmonary bypass.

Keywords: Myxoma; Cardiopulmonary bypass; Leiomyoma; Inferior vena cava

Case Report

A 48 year old woman referred to our department from the department of obstetrics and gynecology who came for the routine checkup after showing a lesion in the right atrium which considered being a blood clot or a primary cardiac tumor. She had the history of total hysterectomy 5 years ago in a local hospital due to a large uterine fibroid. She was fine for the last 5 years after the surgery, but recently she started to have symptoms of breathlessness, swelling of lower limbs, intermittent abdominal pain and giddiness.

On physical examination she was afebrile, conscious, cooperative, and normotensive with slight engorgement of the right jugular vein. Pelvic vascular ultrasound revealed thrombotic features in inferior vena cava and pelvic veins. On further investigation, computed tomography showed a large mass arising from the iliac veins, mainly the left side. It was confirmed that the patient has lesion arising from the pelvic veins and extended via the inferior vena cava up to the right atrium **(Figures 1 and 2)**.

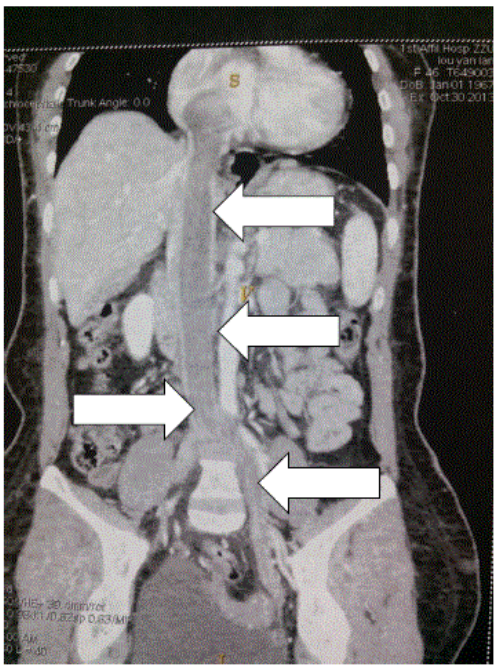

Figure 1: Preoperative computed tomography scan reveals intracaval filling defects extending between the left iliac vein and the right atrium.

After the proper and illustrative diagnosis, it was confirmed that the patient has a leiomyoma with extension to the right atrium. Patient was explained about her condition and the way of treatment. After the consent papers were signed, she was ready for the operation.

The surgical approach was through median sternotomy. The usual process of establishing cardiopulmonary bypass was performed for any emergency purposes of mass bleeding. The aortic cannulation was done and connected to the extracorporeal machine with a single superior venous cannulation into the superior vena cava. The right atrium was palpated and felt the tumor. After the temperature was slightly

decreased to 35°C, cardioplegic solution was pumped and stopped the heartbeat. After complete heart block, the right atrium was opened carefully and felt the tumor directly with the fingers and checked if there were any adhesions on the right atrial wall. After careful inspection and confirming there were no any adhesions, the tumor was held tightly and pulled out slowly to avoid any break in the lower part. As soon as the tumor was totally pulled out with its tail part from the pelvic veins, the other venous cannula was inserted into the inferior vena cava and established complete Cardio Pulmonary Bypass. Right atrium was checked properly for any tearing and bleeding. The patient started rewarming **(Figure 3)**.

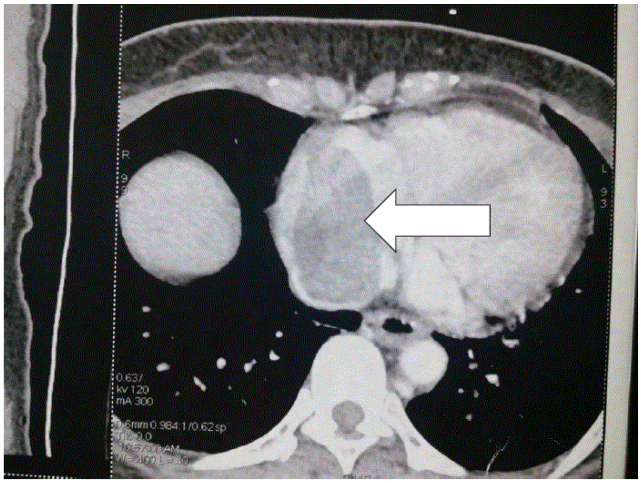

Figure 2: CT showing a mass in the right atrium.

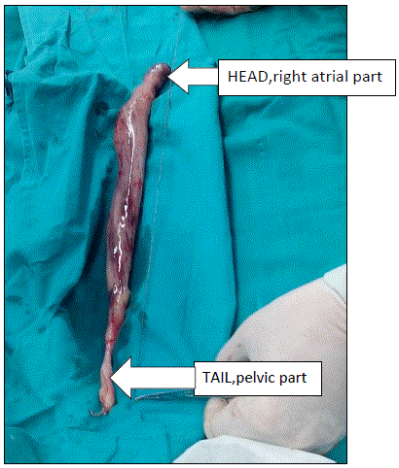

Figure 3: Gross specimen of the tumor removed from the right atrium and inferior vena cava (bottom).

Grossly, the tumor had a white glistening appearance, soft consistency and covered by a true capsule. The length of the tumor was about 45-50cm. After completing the rewarming to the normal temperature, weaned the CPB, closed the chest and sent to the ICU. The post-operative course was uneventful. The patient was discharged on the fifth postoperative day. Pathological examination revealed that the mass composed of benign smooth muscle cells with fibrous tissue consistent with leiomyoma. There was no any evidence of sarcomatous changes. Follow up of the patients in the 3rd month revealed normal and no recurrence of tumor.

Discussion

This case was confirmed as benign leiomyoma extended to the heart based on clinical history, morphological evaluation and histopathological work up. It is also known as intravenous leiomyoma (IVL) extended to heart. IVL is a condition which only affects women, mostly with the previous history of hysterectomy due to uterine fibroid [2].

Birch-Hirschfeld first presented a case of intracardiac extension of IVL in 1896 [3], and Durck first presented a case of itracardiac extension of IVL in 1907, with more than 100 reported cases since early 1900s [4]. Since IVL is a rare case, it is usually not in the mind of paractitioners to include in the differential diagnosis of an intracardiac tumor which may lead to the misdiagnosis of the case. Usually, right atrial mass is detected at first in patients with cardiac symptoms as in our patient. So, we can create a mistake to diagnose it as myxoma if we do not take the proper history, hence it's a clue that any intracardiac tumor in women must contain IVL as a part of differential diagnosis.

In our case, we were able to make a proper diagnosis before surgery due to careful mediacal history and CT. Usually, IVL is diagnosed by echocardiography, CT or MRI [5].

There have been two theories proposed to explain the origin of IVL [6, 7]. One suggests that the tumor arises from the vascular wall, and the other says about the vascular invasion of the myometrial veins by the leiomyoma. But the exact mechanism of origin is still under study.

A total surgical excision is the most successful therapy for IVL. Different ways of treatment have been reported; as one stage and two stage resection therapies, but our patient was little simple because she had already done the hysterectomy, so only one stage complete resection was enough for her treatment. First total resection was reported in 1982 by Ariza et al. [8] by two stage resection therapies with delayed laparotomy after resection of intracardiac part of the tumor.

Conclusion

IVL extended to the heart is a rare case which may be easily misdiagnosed without proper medical history and imaging technique. Abdominal CT is a useful imaging technique for IVL. Also, when a right atrial mass is detected in female with previous history of hysterectomy due to leiomyoma or a present history of uterine fibroid, IVL should be considered. Since the cardiac extension in IVL may be long delayed, we recommend a prolonged follow up.

References

1. Clement PB (1988) Intravenous leiomyomatosis ofthe uterus. Pathology Annual 23: 153-183.
2. Harris LM, Karakousis CP (2000) Intravenous leiomyomatosis with cardiac extension: tumor thrombectomy through an abdominal approach. Journal of Vascular Surgery 31: 1046-1051.
3. Birch-Hirschfeld FV (1896) Lehrbuch der Pathologischen Anatomie. CW Vogel (5th edn.) Leipzig, Germany.
4. Kocica MJ, Vranes MR, Kostic D, Kovacevic-Kostic N, Lackovic V, et al. (2005) Intravenous leiomyomatosis with extension to the heart: rare or underestimated? J Thorac Cardiovasc Surg 130: 1724-1726.
5. Hayasaka K, Tanaka Y, Fujii M, Himi K, Negishi N (2000) Intravenous leiomyomatosis. Journal of Computer Assisted Tomography 24: 83-85.
6. Norris HJ, Parmley T (1975) Mesenchymal tumors of the uterus-Intravenous leiomyomatosis. A clinical and pathologic study of 14 cases. Cancer 36: 2164-2178.
7. Tierney WM, Ehrlich CE, Bailey JC (1980) Intravenous leiomyomatosis of the uterus with extension into the heart. American Journal of Medicine 69: 471-475.
8. Ariza A, Cerra C, Hahn IS, Shaw RK, Rigney B (1982) Intravascular leiomyomatosis of the uterus-A case report. Conn Med 46: 700-703.