Case Report

Nepalese Heart Journal 2020; Vol 17 (2), 47-49

Intravenous Leiomyoma with Intracardiac Extension: A Rare Late Presentation After Hysterectomy.

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Cite this article as: Jaiswal LS, Pandit N, Prasad JN, et al. Intravenous Leiomyoma with Intracardiac Extension: A Rare Late Presentation After Hysterectomy. Nepalese Heart Journal 2020; Vol 17 (2), 47-49

Submitted date: 18th August 2020
Accepted date: 30th August 2020

Abstract

Intravenous leiomyoma is a rare, benign smooth muscle tumor arising from intrauterine venules or the myometrium. It can rarely present as intracardiac mass long after hysterectomy. In this case report we describe a 40-year-old female with previous history of hysterectomy, presenting with an intracardiac mass which was successfully managed with single stage tumor resection under cardiopulmonary bypass. Subsequent histopathology showed features of leiomyoma. The diagnosis of intravenous leiomyoma with cardiac extension should be kept in a female patient presenting with intracardiac mass with previous history of myomectomy or hysterectomy.

Keywords: Intracardiac; Intravenous; Leiomyoma; Tumor.

DOI: https://doi.org/10.3126/njh.v17i2.32680

Introduction

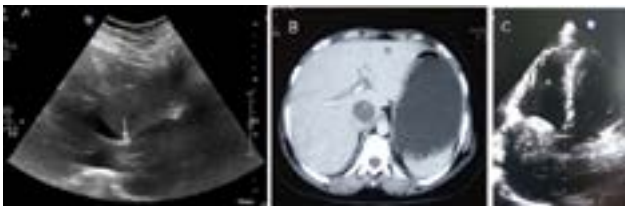
Primary cardiac tumors in general are rare and predominantly benign.[1]Primary cardiac leiomyoma is even rarer[2], most of the cardiac leiomyomas are secondary to tumor metastasis or in continuity extension from the uterine leiomyoma.[3,4] Intravenous leiomyoma is defined as a benign, smooth muscle tumor arising from the intrauterine venules and/or the myometrium with identifiable growth within the lumen of veins.[3] Here we describe a single stage surgical management of a rare intravenous leiomyoma presenting late after hysterectomy as intracardiac mass.

Case Report

A 40-year-old female presented to local physician with 5-months history of pain in the right upper abdomen. She had significant past medical history of hysterectomy for symptomatic uterine leiomyoma eight years back. There was no evidence of congestive heart failure. Her general physical examination and laboratory investigations (liver function test, renal function test and complete blood counts) at presentation were within normal limits. Abdominal ultrasonography, revealed a tumor thrombus in the retrohepatic inferior venacava (IVC) (Figure 1, A). Contrast enhanced computed tomogram of abdomen confirmed the tumor thrombus in the IVC (Figure 1, B)

Extending from the gonadal vein till cavo-atrial junction. Both renal parenchyma were normal in CT. Transthoracic echocardiogram showed a mobile mass extending from the IVC into the right atrium (RA) and right ventricle (RV) (Figure 1, C).

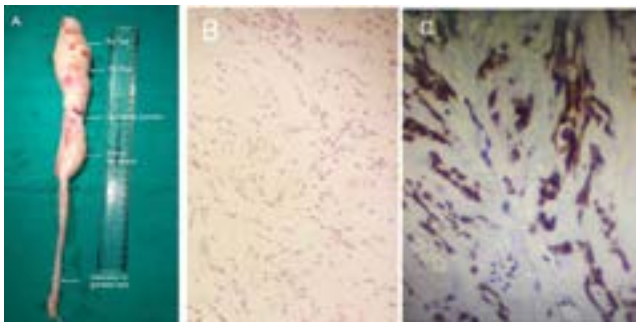
Figure 1.



A. Ultrasound abdomen showing mass in the retrohepatic IVC extending to the RA (arrow at cavoatrial junction);
B. CT abdomen showing mass in the retrohepatic IVC (asterisk);
C. Echocardiography showing mass (asterisk) in the RA protruding into the RV via tricuspid valve.

An operative decision was made, as the mass was large and mobile, almost completely occupying the RA with impending risk of sudden death due to tricuspid valve occlusion and massive pulmonary embolism. The tumor thrombus was approached via median sternotomy under cardiopulmonary bypass (CPB) and moderate hypothermia. After median sternotomy the aorta, superior venacava (SVC) and right femoral vein (conservative length till below infrahepatic IVC) were cannulated, CPB initiated, and the patient was cooled down to 32°C. After opening the RA, intracardiac part of the tumor was seen freely floating, however, it was snuggly fitted at RA-IVC junction that was freed by blunt finger dissection. The entire tumor from cardiac to the infrarenal IVC and gonadal vein extension was easily pulled-out with little tension. The RA was closed and CPB weaned off. Grossly, the tumor was firm, rubbery, grey-white and of 32 cm long and maximum thickness of 5 cm in the RA part (Figure 2, A). Histopathology revealed leiomyoma with spindle to stellate shaped cells without any abnormal mitotic figures (Figure 2, B). Immunohistochemistry showed the spindle cells positive for actin, vimentin and desmin and negative for CD-34 and S-100 (Figure 2, C). The staining for estrogen and progesterone receptors was positive, suggesting uterine origin.

Figure 2.



A.Gross image showing the tumor size and location at different levels;
B.Histopathology (H&E stain) showing spindle to stellate cells and;
C. Immunohistochemistry showing positive stain for desmin

At 18 months of follow-up patient is asymptomatic. Echocardiography and abdominal ultrasonography showed no evidence of recurrence.

Discussion

Three clinical settings have been described for intracardiac leiomyoma: primary cardiac leiomyoma, intravenous leiomyoma with cardiac extension and benign metastasizing leiomyoma.[2] Intravenous leiomyoma is an unusual consequence of pelvic leiomyoma originating either in the uterus or in the walls of the pelvic veins. The attachment site for intravenous leiomyoma is mostly intra-abdominal which includes ovarian or hypogastric veins and their entrance into the iliac vein or the IVC.[5]

It usually presents in females at fourth or fifth decades of life.[2] The presentation of disease varies from asymptomatic and incidentally detected tumor to frank heart failure.[4] It usually has an indolent course and remains dormant in the pelvic vein until detected incidentally, hence present delayed after hysterectomy. It can rarely extend upto the IVC and right atrium, and can present as cardiac failure.[3] On contrary, in our case, there were no symptoms of heart failure despite large size and intracardiac extension. The diagnosis of intravenous leiomyoma with cardiac extension should be kept in a female patient presenting with intracardiac mass with previous history of myomectomy or hysterectomy for leiomyoma.

Successful treatment involves in toto excision, including both abdominal and thoracic part of the tumor. Several approaches have been described for complete removal of the intravenous leiomyoma with intracardiac extension. The tumor can be removed in a staged manner, which involves removal of thoracic part tumor under cardiopulmonary bypass followed by laparotomy for the abdominal part. It can also be removed in single setting via combined median sternotomy and midline laparotomy approach under CPB with or without deep hypothermic circulatory arrest (DHCA). Single stage abdominal approach without cardiopulmonary bypass has also been described.[5] We initially planned for single stage removal via median sternotomy and midline laparotomy. However, intraoperative transesophageal echocardiogram (TEE) showed the mass to be freely floating in both IVC and intracardiac part; furthermore superficial inspection and palpation of tumor over RA and IVC confirmed it to be purely intraluminal firm mass without any infiltration to IVC. We were able to successfully remove the entire tumor in single stage via median sternotomy approach without the need for laparotomy and DHCA. Postoperative TEE and abdominal ultrasound confirmed the complete removal of the tumor.

Conclusion

Intravenous leiomyoma should be kept in differential diagnosis of intracardiac mass, especially in middle age women with previous history of hysterectomy. It can be successfully treated with surgical excision.

Acknowledgement: None

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