Three-Dimensional Echocardiographic Evaluation of Intravenous Leiomyomatosis: Case Report

İntravenöz Leomiyomatozis Olgusunun Üç Boyutlu Ekokardiyografi ile Değerlendirilmesi

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ABSTRACT Intravenous leiomyomatosis is a rarely-seen, benign tumoral mass originated from smooth muscle cell of either the uterine venous wall or uterine leiomyoma and sometimes may spread into the venous cavities and right heart chamber. It usually affects premenopausal women. Despite its benign histology, cardiac involvement is a serious complication. The choice treatment is surgery with complete tumour resection. In this case report, we present a patient with intravenous leiomyomatosis with cardiac extension and discuss the diagnosis and treatment approaches.

Anahtar Kelimeler: Ekokardiyografi; ekokardiyografi, 3 boyutlu; kalp tümörleri

ÖZET İntravenöz leomiyomatozis, uterin venöz duvar ya da uterin leomiyom düz kasından köken alan nadir ve iyi huylu tümoral bir kitledir. Bazı durumlarda venöz boşlukta ilerleyerek sağ kalp boşluğuna kadar ulaşabilir. Sıklıkla premenopozal dönemdeki kadınlarda görülür. İyi huylu histolojik yapısına rağmen kardiyak tutulum ciddi bir komplikasyondur. Tedavide tümörün tamamının çıkarılması gerekir. Bu olgu sunumunda kardiyak tutulumu olan intravenöz leomiyomatozis tanılı hasta sunulmakta ve tanı ile tedavi yaklaşımı tartışılmaktadır.

Key Words: Echocardiography; echocardiography, three-dimensional; heart neoplasms

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Intravenous leiomyomatosis is a rare tumor characterized by benign histology.[1] Although leiomyoma is generally limited to the uterus, it sometimes extends into the inferior vena cava and reaches to right side of the heart by infiltrating uterine or ovarian veins. In this report, we present a female patient with intravenous leiomyomatosis extending to the right side of the heart evaluated by real time three dimensional (3D) echocardiography.

CASE REPORT

A 43-year-old female patient who had been evaluated for a right atrial mass on echocardiography referred to our hospital with presumptive diagnosis of atrial myxoma. Transthoracic echocardiography (TTE) showed a right atrial mass which was moving to right ventricle at diastole (Figure 1A). At subcostal view, inferior vena cava (IVC) was dilated and the mass was found to

be extending from IVC to the right atrium (Figure 1B). Transthoracic real time 3D echocardiography imaging by using Vivid 7 Dimension echocardiography equipment (GE, Vingmed, Horten, Norway) was performed and revealed that the lobulated mass had a homogenous appearance with a smooth surface. No protrusion above the mass surface or pedicle in relation the interatrial septum was present (Figure 1C). The mass was found to be infiltrating the IVC wall (Figure 1D). With the aid of abdominal ultrasonography and computed tomography, a heterogeneous mass (13<7 cm in size) extending superiorly from pelvic entrance along the IVC to the right atrium which is compatible with leiomyomatosis was diagnosed. She was operated on with two-stage approach to excise the mass. At the first stage, the intracardiac part of the mass was resected with right atriotomy (Figure 2). One month later, the residual vascular part of the mass was resected and total hysterectomy and bilateral salpingo-oopherectomy were performed. Histological examination confirmed the diagnosis of intravascular and intracardiac leiomyomatosis.

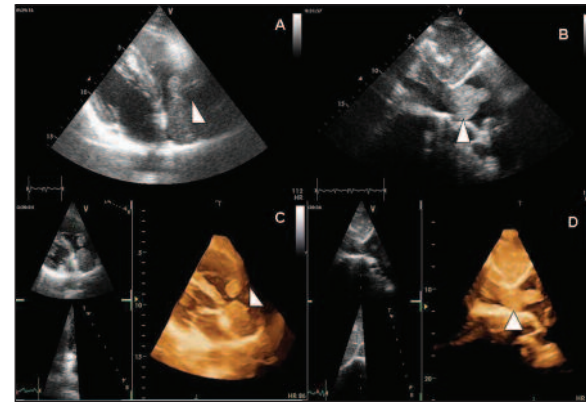

FIGURE 1A-D: A) Transthoracic echocardiograpy showing a right atrial mass **B)** Transthoracic echocardiograpy from the subcostal view showing extension of the mass from inferior vena cava to the right atrium. **C)** Transthoracic 3D echocardiography showing the mass with a smooth surface. **D)** Transthoracic 3D echocardiography showing tumoral infiltration of the inferior vena cava wall.

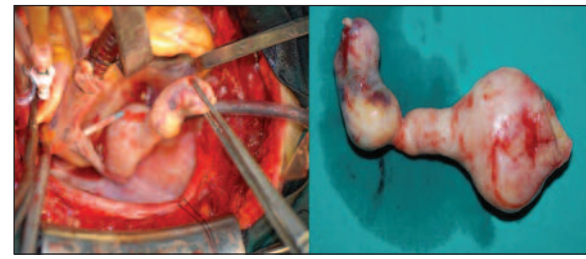

FIGURE 2: Macroscopic view of resected mass.

DISCUSSION

Leiomyomatosis was first described by Birch-Hirschfeld as a rarely seen, benign tumoral mass originated from smooth muscle cell.[2] This entity may originate from either the uterine venous wall or uterine leiomyoma and spread into the venous cavities and right heart chambers.[3,4] Benign and malign tumors, thrombus and tumor thrombus should be kept in mind for differential diagnosis of right atrial masses. Other than intravenous leiomyomatosis, extracardiac tumors such as renal cell carcinoma, hepatocellular carcinoma, Wilms tumor and adrenal tumor may present with similar clinical profile. Extracardiac tumors usually reach to the right atrium by the involvement of inferior vena cava.[5]

As an easy, reproducible and cheap diagnostic method, transthoracic echocardiography is the first choice in determining and evaluation of intracardiac massses. Besides, evaluation of IVC from subcostal view may show the involvement of IVC and may be a clue for the origin of the mass. Frequently, cases of intravenous leiomyomatosis have been misdiagnosed as myxoma or atrial thrombus.[6] Myxoma usually arises from the fossa ovalis of the interatrial septum at left atrium; however it may be also localized to any other cardiac chamber or valves. Presence of a narrow base attachment may be helpful for the diagnosis of myxoma.[7] The differentiation between tumor and thrombus is not always possible with TTE. As compared with the 2D echocardiography, real time 3D echocardiography is a new diagnostic tool that can provide further information about cardiac masses with regard to its size, structure and the relation to the other cardiac structures with a superior spatial resolution.[8] In addition, 3D echocardiography is able to show whether intracardiac masses are free floating or attached to the wall of a cardiac chamber.[9] This feature may particularly be useful in differentiating between tumor and thrombus. In the present

case, 3D echocardiography was performed to further delineate the mass and its relation to other cardiac structures, and revealed a free floating mass with a smooth surface. These features suggested a tumoral origin of the mass rather than a thrombus formation. To the best of our knowledge, this is the first demonstration of intravenous leiomyomatosis with 3D echocardiography in the literature so far.

In summary; in a middle-aged female patient, especially with the history of hysterectomy or the diagnosis of myoma uteri, intravenous leiomyomatosis should be kept in mind in differential diagnosis of right atrial masses. A new imaging tool, 3D echocardiography, may provide additional information for differential diagnosis and management of this rare clinical entity.

REFERENCES

1. Norris HJ, Parmley T. Mesenchymal tumors of the uterus. V. Intravenous leiomyomatosis. A clinical and pathologic study of 14 cases. Cancer 1975;36(6):2164-78.
2. Arinami Y, Kodama S, Kase H, Tanaka K, Okazaki H, Maruyama Y. Successful one-stage complete removal of an entire intravenous leiomyomatosis in the heart, vena cava, and uterus. Gynecol Oncol 1997;64(3): 547-50.
3. Nam MS, Jeon MJ, Kim YT, Kim JW, Park KH, Hong YS. Pelvic leiomyomatosis with intracaval and intracardiac extension: a case report and review of the literature. Gynecol Oncol 2003;89(1):175-80.
4. Kılıç SH, Alagöz A, Tuncay G, Akbay SI, Mollamahmutoğlu L. [Leiomyomatosis peritonealis disseminata associated with pregnancy: case report]. Turkiye Klinikleri J Gynecol Obst 2005; 15(4):214-16.
5. Nakayama DK, Norkool P, deLorimier AA, O'Neill JA Jr, D'Angio GJ. Intracardiac extension of Wilms' tumor. A report of the National Wilms' Tumor Study. Ann Surg 1986;204 (6): 693-7.
6. Topcuoglu MS, Yaliniz H, Poyrazoglu H, Tokcan A, Demir SC, Bozkurt A, et al. Intravenous leiomyomatosis extending into the right ventricle after subtotal hysterectomy. Ann Thorac Surg 2004;78(1):330-2.
7. Araoz PA, Mulvagh SL, Tazelaar HD, Julsrud PR, Breen JF. CT and MR imaging of benign primary cardiac neoplasms with echocardiographic correlation. Radiographics 2000;20(5): 1303-19.
8. Asch FM, Bieganski SP, Panza JA, Weissman NJ. Real-time 3-dimensional echocardiography evaluation of intracardiac masses. Echocardiography 2006;23(3):218-24.
9. Upendram S, Nanda NC, Mehmood F, Vengala S, Dod HS, Bodiwala KN, et al. Images in geriatric cardiology: live three-dimensional transthoracic echocardiographic assessment of right atrial thrombus. Am J Geriatr Cardiol 2004;13(6):330-1.