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Intravenous Leiomyomatosis with Right Atrial Involvement

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SAĞ ATRİYUMU TUTAN İNTRAVENÖZ LEİOMİYOMATOZ

Intravenöz leyomiyomatoz genellikle uterus miyomundan kaynaklanan nadir bir neoplastik hastalık olarak kabul edilmektedir. Bu hastalıktan ölüm genellikle kalp tutulumu sonucudur.

Bu yazıda uterus miyomu nedeni ile histerektomi geçirmiş 42 yaşında bir kadın hastada inferior vena kava ve sağ atriyuma yayılan intravenöz leyomiyomatoz bildirilmektedir.

Histolojik olarak selim olan tümör sağ atriyum ve inferior vena kavanın suprahepatik bölümünden çıkarılmıştı. Her nekadar bunu izleyen laparatomide, infrarenal vena kava inferior ve sağa vena iliaka kommunisi tutan tümör yapışıklıklar nedeniyle tam olarak çıkarılmamış ise de, hasta 14 aylık izleme boyunca asemptomatik kalmıştır.

Anahtar kelime: *Intravenöz leyomiyomotoz, sağ atriyum, vena kava inferior*

Intravenous leiomyoma is a rare, benign, smooth-muscle tumor that originates mostly from the uterus and can extend through the pelvic or ovarian veins to the inferior vena cava (IVC) and right atrium. The inferior venacavogram and tomographic scan have proved to be extremely helpful in the diagnosis and management of IVC tumors with right atrial involvement. We present a patient with a preoperative diagnosis of right atrial myxoma and IVC thrombosis that ultimately proved to be an intravenous leiomyoma.

CASE REPORT

A 42-year-old woman, complaining of dyspnea and palpitation, was referred to the Department of Cardiovascular Surgery, Gülhane Military Medical Academy, with the preoperative diagnosis of intracardiac myxoma complicated with thrombosis in the IVC. She had 3 normal deliveries and had a total abdominal hysterectomy at age 37 for benign uterine myoma. On physical examination blood pressure was 135/75 mm Hg, pulse rate was 108 beats per minute, a 2/6 systolic murmur was heard at the lower right sternal border, the liver was palpable 2 cm below the costal margin, and minimal ankle edema was dedected. The chest X-ray and the routine laboratory values were normal. The electrocardiogram showed sinus tachycardia and nonspecific T wave inversions. A transthoracic two-dimensional echocardiogram revealed a 6x8 cm right atrial mass extending distally into the IVC. The mass prolapsed through the tricuspid valve into the right ventricle during diastole. The inferior venacavogram showed an extensive filling defect of IVC with large collaterals towards the azygos and paravertebral veins (Fig. 1). The patient was subjected to surgery for removal of the cardiac mass and IVC thrombosis. Operation was performed through median sternotomy. Cardiopulmonary bypass was instituted through the right atrial appendage and the ascending aorta. During core cooling to 20 °C and circulatory arrest, the right atrium was opened. A large, yellowish, solid, elastic-soft tumor, extending from the IVC into the right atrium was found. It was free lying and without evidence of infiltration into the right atrium, but there were extensive adhesions at the IVC component beyond the level of the proximal hepatic veins. The intraatrial and suprahepatic IVC tumor was excised completely (Fig 2). Hepatic veins were seen to be draining freely into proximal IVC. Hepatic venous drainage was controlled with cardiotomy suction. No attempt was made to excise the tumor in the intrahepatic IVC at that time because of extensive adhesions. Cardiopulmonary bypass was reinstituted after 12 minutes of circulatory arrest and the operation was completed in routine fashion. The intraoperative frozen-section diagnosis was benign leiomyoma. The patient made a good recovery postoperatively. A laparotomy was performed a month later. The infrarenal IVC was found to be grossly distended with tumor. The right common iliac vein contained tumor, but the left iliac vein was unaffected. No abdominal mass was found. IVC was opened separately below the level of the renal veins and the right common iliac vein. At these levels, the tumor was found to be moderately adhesive to the posterior wall of the IVC and was removed from this site, using cutting wire loop. However, there was extensive adhesion in the suprarenal IVC, and the tumor could not be removed between the infrahepatic and suprarenal portions of IVC. The right common iliac vein was clipped at the level of the bifurcation to retard subsequent propagation of the tumor into the IVC. Although intravascular tumor was left above the renal veins, all the remaining (about 75%) tumor mass was extracted from the IVC and the heart. His-

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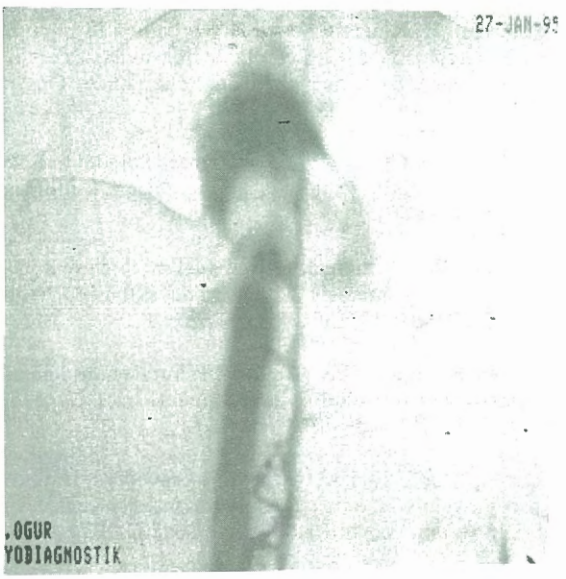

Fig. 1: Inferior venacavogram demonstrating extensive filling defect of right atrium and inferior vena cava, and large collateral veins.

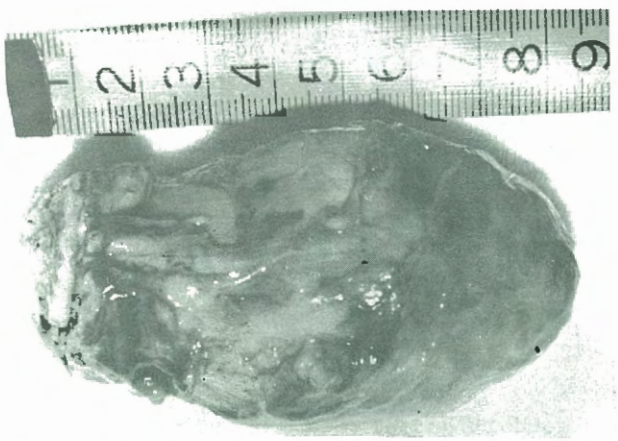

Fig. 2: The tumor after excision from the heart and suprahepatic vena cava inferior.

tological examination of tumor tissue from both operations confirmed the diagnosis of intravenous leiomyoma. The patient had an uneventful postoperative course and was treated by anticoagulation for risk of IVC thrombosis and pulmonary emboli. At the 14th month following discharge, she was asymptomatic and had a normal physical examination. Postoperative inferior venacavogram showed no filling defect and no collaterals (Fig 3).

DISCUSSION

Intravenous leiomyomatosis has been defined as a rare neoplastic disease characterized by extension into venous channels of histologically benign smooth muscle tumors arising from a uterine vessel [1]. It produces various clinical presentations, such as right-sided cardiac failure, dyspnea, and palpitation,

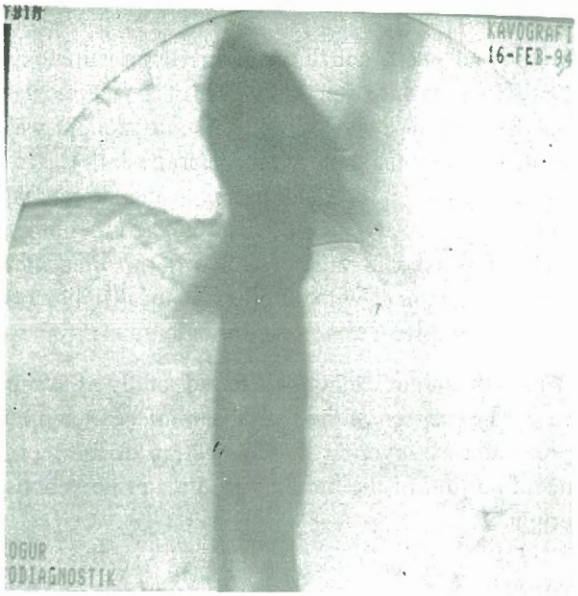

Fig. 3: Postoperative inferior venacavogram showing filling defect only in midportion of the inferior vena cava.

attributable to impairment of the tricuspid valve [2]. Other unusual features are cerebral embolization and Budd-Chiari syndrome [3]. Symptomatology varies according to the location. All reported patients are female and over 90 % of them are premenopausal and parous [4,5]. A history of previous hysterectomy or pelvic surgery was present in about 10 % of these patients [5].

Clinical diagnosis of intravenous leiomyomatosis with cardiac involvement remains difficult. Modern imaging techniques such as echocardiography, tomographic scanning (CT), magnetic resonance imaging (MRI) and angiography have enabled an increasing number of cases to be diagnosed preoperatively [3]. CT, MRI, and inferior venacavogram demonstrates tumors and invated areas in IVC. Imaging techniques are important in ensuring early and accurate definition of these tumors, so as to aid planning of surgical management. The prognosis of intravenous leiomyomatosis is usually reported as favorable [3-7]. Surgery appears to be the only treatment that has significant effect on survival.

In our patient, preoperative diagnosis was right atrial myxoma complicated by thrombosis in the IVC. Diagnosis of intravenous leiomyomatosis with cardiac involvement should be considered in a female patient presenting with signs of cardiac myxoma with a history of hysterectomy. In our patient, there was no

abdominal mass, and intravenous leiomyomatosis had arisen from the smooth muscle of the uterine veins. Because of extensive adhesion, no attempt was made to excise the tumor in the suprarenal IVC. However, about 75 % of the tumor in the suprarenal IVC was evacuated and the continuity of the tumor was obolished. Our patient is alive 14 mounths after surgical treatment without signs of local tumor recurrence or distant tumor manifestations.

If the IVC tumor cannot be excised totally as in our case, re-extension of the tumor into the heart can be prevented by severing the connection of the more distal portion of the tumor from its uterine venous origin.

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