Severely Calcified Intravenous Leiomyomatosis with Cardiac Extension

Intravenous leiomyomatosis (IVL) is a rare benign tumor originating from the uterus, and the tumors of this type with intracardiac extension are even rarer. In this article, we report the case of an older patient with IVL that spread into the heart from the left internal iliac vein. Several atypical features of this case compared with other reports are described.

Case

A 72-year-old female was referred to our hospital due to syncope. She had a history of hysterectomy for uterine myoma 30 years ago, the details of which were not available at admission. Before the onset of her syncope, she had been in excellent health. Upon admission, her blood pressure was 153/72 mmHg and her heart rate was 76 beats per minute. A systolic murmur was audible at the margin of the fourth intercostal space on the left sternal border. The laboratory values were normal. Echocardiogram was performed which revealed an intracardiac tumor in the right atrium (RA) with an acoustic shadow that arose from the inferior vena cava (IVC) (Fig. 1). The tumor head entered the right ventricle (RV) through the tricuspid valve during diastole and retracted into the RA during systole. Computed tomography revealed a severely calcified tumor extending from the left internal iliac vein to the RA (Fig. 2). A subsequent inferior vena cavaogram also demonstrated an intraluminal filling defect from the internal iliac vein to the RA (Fig. 1). A subsequent inferior vena cavaogram also demonstrated an intraluminal filling defect from the internal iliac vein to the RA. The tumor was attached to the left common and internal iliac vein and floated freely in the caval lumen (Figs. 3, 4). A simultaneous operation was performed with a median sternotomy and left retroperitoneal approach to the left iliac vein. Cardiopulmonary bypass was instituted using cannulas through the right femoral vein, the superior vena cava, and the ascending aorta. The left internal iliac vein was incised and the stalk of the tumor in the vein was excised. Despite these efforts, a part of the tumor remained in the distal internal iliac vein. The body of the tumor still adhered to the common iliac vein. Then, the RA was opened during core cooling to 28°C with 30 seconds of circulatory arrest. There were no vis-
Fig. 1. Echocardiogram revealed the tumor in the right atrium that arose from the inferior vena cava. RA, Right atrium; IVC, inferior vena cava.

Fig. 2. Computed tomography shows the tumor with calcification in the right atrium.

Fig. 3. Venography revealed an intraluminal filling defect from the left internal iliac vein to the right atrium (arrows).

Fig. 4. Schema of the tumor and venous system. The tumor head entered the right ventricle during diastole and retracted into the right atrium during systole. The tumor was only attached to the left common and internal iliac vein. RA, Right atrium; RV, right ventricle; IVC, inferior vena cava; CIV, common iliac vein; IIV, internal iliac vein.