A 15-year-old girl of Middle East origin was seen by a pediatrician because of suspected pituitary nanismus. That diagnosis was quickly ruled out; however, a supposedly innocent systolic cardiac murmur resulted in referral to echocardiography. She had no cardiopulmonary symptoms, but the examination revealed a large nonobstructive, mobile, and apparently multicystic mass in the right ventricle (Figure 1). The mass was located close to the tricuspid valve with a stalk to a papillary muscle. Color Doppler imaging did not show any flow in the tumor (Figure 2; also see online-only Data Supplement Movie clips).

A substantial diagnostic workup was initiated. The patient often visited her family in the Middle East, and this in combination with the multicystic appearance of the mass led to a tentative diagnosis of hydatid cyst, caused by echinococcosis, although a cardiac tumor was obviously also considered.1 All blood tests were normal including her infectious parameters, and she did not have serum antibodies to any Echinococcus species. An FDG PET-CT scan did not show any particular FDG activity in the mass. Cardiac magnetic resonance showed no signs of tumor infiltration into the myocardium, a feature supporting the suggestion of a benign tumor (Figure 3). It was decided to perform a flash-contrast echocardiography to elucidate the vascularity of the mass and the cyst-like structures (Figure 4). For the contrast echocardiography, a Vivid 7 (GE, Horten, Norway) ultrasound scanner was used, equipped with a 2.5-MHz probe. The echocastagent (Sonovue, Bracco, Inc, Milan, Italy; diluted in 10 mL isotonic sodium chloride) was administered intravenously with isotonic sodium chloride (approximately 250 mL/h) at constant rate (approximately 1 mL/min), which kept the contrast dissolved during the infusion. The scanner was adjusted to a low mechanical index to diminish contrast disruption, followed by short bursts of high mechanical index to destroy the contrast in the field of view, including the tumor (Figure 3B). During 15 consecutive heart beats, the perfusion in the tumor was semiquantitatively assessed by comparing contrast replenishment in the interventricular septum with contrast replenishment in the tumor.

This examination unveiled that in reality, the apparent cysts in the mass represented irregular and dilated vascular formations. Accordingly, the cyst-like processes represented large venous sinusoids.

At open heart operation, the tumor was completely extirpated (Figure 5A). It measured 20×20×30 mm and was polypoid, though smoothly outlined. The microscopy was typical for a myxoma but also revealed large vessels stretching from the periphery of the tumor all the way to its base (Figure 5B), a finding in accordance with the flash-contrast echocardiography.

Figure 1. Echocardiography. A, Apical 5-chamber view showing the attachment of the mass (M) to a papillary muscle in the right ventricle. B and C are parasternal long-axis and short-axis views, respectively. In these views, the multicystic appearance of the mass is clearly seen. AO indicates aorta; LA, left atrium; LV, left ventricle; and RA, right atrium.
In summary, myxomas are very rare in adolescents, in particular in the ventricles, and the most common presentation is a solid tumor. Yet, the distinction between cystic cardiac tumor and hydatid cyst using color Doppler echocardiography has been reported previously. Using varying pulse repetition frequencies in the present case, we could not detect the very slow flow in the venous sinusoids by means of color Doppler echocardiography.

The use of contrast echocardiography is generally recommended in cases with intracardiac mass lesions and helps distinguish thrombi without perfusion from tumors with varying degrees of perfusion, depending on tumor origin. In the present case, flash-contrast echocardiography documented the apparent cysts to be vascular and thus supplemented the information obtained from the more advanced imaging modalities.

However, it should be stressed that apart from the microscopy, none of the multiple imaging techniques could provide a reliable differentiation between various types of tumor pathologies such as rhabdomyoma, sarcoma, teratoma, or myxoma, an issue of great importance for prognosis and postoperative care.

Disclosures

None.

References


Key Words: cystic mass / echocardiography / tumor / women

Figure 2. Echocardiographic image with color Doppler. The velocity scale is adjusted to 49 cm/s. There is no flow in the cardiac mass. Lower color flow velocities gave aliasing artifacts in the tumor and the surrounding structures.

Figure 3. Cardiac magnetic resonance images in 4-chamber view showing a large noninfiltrating mobile mass (M), attached to a papillary muscle in the right ventricle. A, On T1-weighted spin-echo, the mass has similar signal intensity as the myocardium. B, By means of T2-weighted imaging, the mass appears with hyperintense signal intensity. LA indicates left atrium; LV left ventricle; and RV, right ventricle.
Figure 4. Flash-contrast echocardiography. Serial images in parasternal long-axis view. A, Continuous pump-mediated intravenous infusion of contrast medium (Optison, GE Health Care) provides constant opacification of the right and left ventricles (RV and LV, respectively) and of the myocardium and the mass (M). The high-power ultrasound flash (B) destroys the microbubbles in the mass and myocardium, as illustrated in C. In D and E, replenishment of the mass reveals that the multiple cysts in the mass represent vascular sinusoids. F, Subsequently, the tissue of mass again is more diffusely filled with contrast medium.

Figure 5. A, The extirpated myxoma. B, Microscopic section through the myxoma (Masson trichrome stain) showing the myxomatous ground substance, at the top lined with endothelium toward the right ventricular cavity. The prominent sinusoids (S) are also lined with endothelium documenting their vascular nature.
Get Closer to the Diagnosis in a Flash
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