Different deformation patterns in intracardiac tumors

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Received 9 September 2004; accepted 12 February 2005
Available online 2 April 2005

KEYWORDS
Cardiac tumors; Strain rate; Echocardiography

Abstract We report on the ultrasound characteristics of two different intracardiac tumors in newborns. The first infant presented with multiple rhabdomyomas in the context of tuberous sclerosis. Ultrasound-based strain and strain rate analysis showed that the tumors deformed in the opposite direction from the surrounding myocardial segments. The second child presented with a large cardiac fibroma as confirmed by pathological examination. In this case strain and strain rate analysis showed the absence of deformation in the tumor. These findings suggest that this new technique could be used to differentiate between different types of myocardial tumors.

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Introduction

Cardiac tumors are rare disorders which can be detected in up to 0.3% of children presenting in congenital heart centers. They are usually detected during infancy and their increased incidence can probably be explained by the availability of better diagnostic tools. The more widespread use of fetal echocardiography has resulted in more tumors being detected during fetal life. Echocardiography has emerged as the current primary diagnostic modality. Its use allows delineation of tumor location and extent, tissue characterization, hemodynamic significance and associated pericardial effusion. Rhabdomyoma and fibroma are the two most common tumors which can be detected in infants. The cardiac rhabdomyoma is a benign tumor of cardiac myocytes. It is composed of altered myocytes with large vacuoles containing large amounts of glycogen. They often occur in association with the tuberous sclerosis complex in which case they appear as multiple masses within...
the heart. The rhabdomyomas have a tendency to spontaneously regress during the first years of life. Treatment is only required when outflow tract obstruction is present.

The cardiac fibroma is a congenital tumor that is composed of fibroblasts and collagen. It is usually solitary and often involves the left ventricular free wall or septum. Tissue characteristics are different between a cardiac fibroma and a rhabdomyoma. In general, a combination of magnetic resonance T1 and T2 sequences allows a description of the composition of the cardiac masses; nevertheless, the initial hope that the pattern of signal intensity would allow differentiation between different types of masses has never been fulfilled. Magnetic resonance myocardial tagging, in addition, can quantify the deformation of these masses and the impact of them on cardiac dynamics. We believe that the different tissue characteristics might cause different deformation patterns within the masses: a rhabdomyoma is softer and contains some sarcomeric elements while a fibroma is a very hard mass which is difficult to deform. A precise assessment of regional deformation might help to better characterize the histology and differentiate tumor tissue from normal adjacent myocardium using echocardiography. This hypothesis was evaluated in two infants with two different types of intracardiac masses. Ultrasound-based strain rate (SR) and strain (ε) imaging was used to evaluate the deformation characteristics of the two different masses.

Case reports

Case 1

In the first baby, a prenatal diagnosis of multiple cardiac tumors, presumably rhabdomyomas, in association with tuberous sclerosis, was made. An uneventful delivery occurred. Three days after the birth an echocardiographic examination showed three tumors within the left ventricle, two of them being attached to the anterolateral wall; the third of them being much larger (18 × 14 × 13 mm). It was attached to the mid segment of the interventricular septum. It did not produce left ventricular inflow or outflow tract obstruction (Fig. 1).

Using color Doppler myocardial imaging, tissue velocity data in three cardiac cycles were recorded in the interventricular septum at high frame rates (>150 frames/s). These velocity data were used for offline analysis using dedicated software (Speqle®, Catholic University of Leuven, Belgium). Regional SR and ε were calculated from the color Doppler velocity dataset by determining the local spatial velocity gradient as previously described. These measurements (Fig. 2) revealed that the structurally normal mid interventricular septal segment had normal deformation (peak systolic SR: 3.7 s⁻¹, peak systolic ε: 54.8%). The rhabdomyoma showed a marked change in shape and volume during the cardiac cycle. It deformed as well, at the beginning of systole in the same direction the myocardium did. However, as soon as

Figure 1  Apical four-chamber view, showing three cardiac tumors within the left ventricle (arrows).
the myocardium started developing contractile forces, it compressed and displaced the mass towards the base of the heart. This led to the tumor being deformed in the opposite direction of the normal surrounding myocardium did (peak systolic SR: $-3.5\, \text{s}^{-1}$, peak systolic $e$: $-43.2\%$).

**Case 2**

A completely asymptomatic newborn was referred for echocardiographic examination after the detection of a systolic murmur localized at the left upper sternal border. During the echocardiographic examination a large mass (19×23 mm) was detected which localized in the right ventricular (RV) free wall extending towards the right ventricular outflow tract. It produced RV outflow tract obstruction with an initial peak gradient of 22 mmHg (Fig. 3). The color Doppler myocardial imaging analysis revealed that the tumor neither moved nor deformed during the different cardiac phases (Fig. 4). Because the RVOT gradient increased during the follow-up, surgical resection of the mass was performed after three months. Histology showed the tumor to be a fibroma.

**Discussion**

These two case reports show that two different types of cardiac tumors have different deformation properties. Rhabdomyomas are deforming in the opposite direction from the surrounding myocardium while fibromas show no deformation. Rhabdomyomas are composed of altered myocytes, which generates a relatively elastic tissue. In our case the mass is squeezed by the surrounding myocardium: it is compressed and displaced towards the base of the heart during systole.
Fibromas consist of noncompliant connective tissue. They are widely and firmly attached to the adjacent myocardial wall. They seem uncompres- sible and do not deform in any direction. Another plausible explanation for the lack of deformation seen in the fibroma may lie in the fact that this tumor was located in the RV which deals with a much lower afterload than the LV; therefore, the squeezing effect, due to high systolic pressure, noted when the mass is located in the LV will not be present when the tumor is situated in the RV free wall.

Non-invasive quantification of regional myocardial deformation properties by ultrasonic SR/3 has made possible to study the relation between regional morphological abnormalities within the ventricles in different cardiac disorders.6–8 The deformation of cardiac tumors has been previously studied by magnetic resonance myocardial tagging; this technique can determine the deformation pattern of a mass precisely.9 Tagging analysis is, however, a very labour intensive technique which is not readily available in most cardiac centers.4

We believe that ultrasound-based deformation imaging could potentially be useful in tissue characterization. Our two cases suggest a potential use of this new technology. Despite these encouraging results, further clinical studies with a larger number of patients and diverse tumors are needed to determine the precise clinical role of ultrasound-based deformation imaging in the extremely rare group of patients with cardiac tumors.

References