


Characterizing a rare cardiac desmoid fibromatosis with multi-modality imaging techniques in a child

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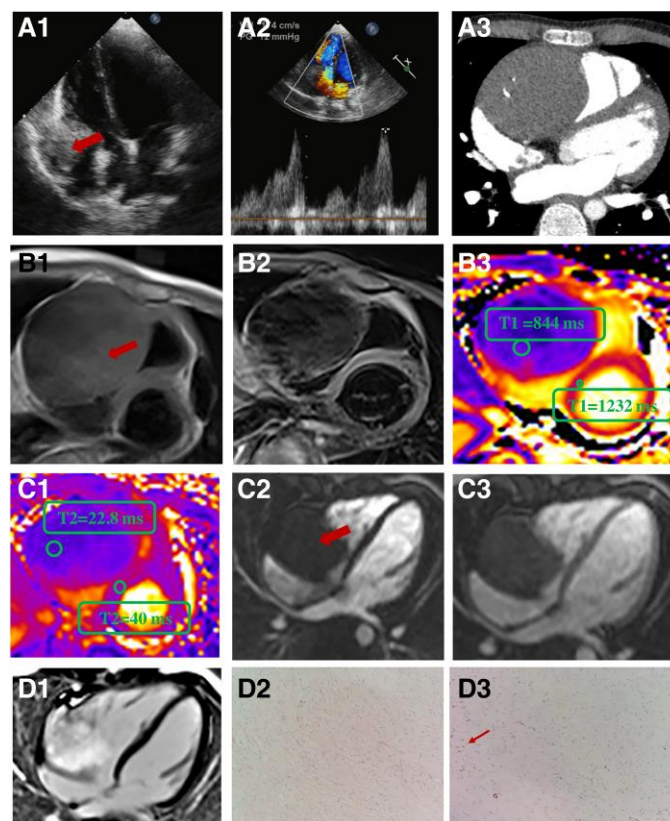


Figure 1 Images of multi-modality imaging techniques and pathology. (A1, A2) Two-dimensional and colour Doppler echocardiographic images; (A3) enhanced computed tomographic images; (B1) T1 weight image; (B2) T2 weight image with fat suppress; (B3) native T1 mapping image; (C1) T2 mapping image; (C2, C3) dynamic perfusion images; (D1) late gadolinium enhancement image; (D2) HE staining images; (D3) immunohistochemical staining of SMA.

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A cardiac mass in a 9-year-old boy was found incidentally while being evaluated for trauma using chest computed tomography (CT). Subsequent transthoracic echocardiography depicted an iso-echogenic mass (6.5 × 5.7 cm) with clear boundary and patchy calcification at free wall of right ventricle (Figure 1A1), and blood flow at tricuspid valve was accelerated (Figure 1A2). Computed tomography angiography showed that the mass presented as soft tissue attenuation with focal calcification and no enhancement was presented within 2 min after contrast agent injection (Figure 1A3), indicating that hemangioma, teratoma, and pleuro-pericardial cyst were excluded. Multi-sequences cardiac magnetic resonance (CMR) was performed to characterize the mass furtherly. Biventricular systolic function was normal with cine images. Compared with myocardium, the mass presented as isointensity in T1 weight sequence (Figure 1B1) and significantly hypointensity in T2 weight sequence (Figure 1B2). Native T1 and T2 values of the mass were significantly lower than that of myocardium in pre-T1 mapping (Figure 1B3) and T2 mapping sequence (Figure 1C1). Slight enhancement was presented in dynamic perfusion sequence (Figure 1C2 and C3). After 5 min of perfusion, the mass presented as homogeneous and obvious late gadolinium enhancement (Figure 1D1). All of these characters on CMR indicated a dense tissue composition and low water content. Thus, fibrous originate mass was suspected. No other organs were involved, and surgery was performed without chemotherapy or radiotherapy. HE staining suspected a mesenchymal tumour (Figure 1D2), and immunohistochemical analysis confirmed a desmoid

fibromatosis with positive smooth muscle actin (SMA) (Figure 1D3). The boy was followed-up regularly at outpatients department without relapse. Primary paediatric cardiac tumours were rare, and most of them were benign. Arrhythmias, blood flow obstruction, and ventricular dysfunction caused by them could result in significant morbidity and mortality. Imaging characters of cardiac tumour on CMR are more predictive of histological diagnosis than CT and echocardiography. Sequences and characteristics of different paediatric cardiac mass were concluded in expert consensus on paediatric CMR.¹ We herein reported this case to highlight the value of CMR for characterizing histological features of paediatric cardiac mass, which is of paramount importance for treatment strategy decision.

Consent: Consent form was signed and obtained from the patient and patient's parents in accordance with COPE guidelines for the publication of the case report.

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Reference

1. Dorfman AL, Geva T, Samyn MM, Greil G, Krishnamurthy R, Messroghli D, Festa P, Secinaro A, Soriano B, Taylor A, Taylor MD, Botnar RM, Lai VWW. SCMR expert consensus statement for cardiovascular magnetic resonance of acquired and non-structural pediatric heart disease. *J Cardiovasc Magn Reson* 2022;**24**:44.