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Images in Congenital Cardiac Disease

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Neonatal right ventricular mass presenting with pericardial effusion and tamponade

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Abstract

Prenatally diagnosed cardiac mass in a newborn caused cardiac tamponade. Cardiac MRI and histopathology confirmed a rare right ventricular mesenchymal hamartoma, previously reported only in the left ventricle.

A male newborn prenatally diagnosed with a cardiac mass presented with cardiac tamponade. He required repeated pericardiocentesis after echocardiography demonstrated a severe pericardial effusion (Video 1). Cardiac magnetic resonance imaging revealed a large subepicardial lesion at the right ventricular apex (Video 2). The mass was isointense to the myocardium on T1-weighted double-inversion recovery images and hyperintense on T2-weighted triple-inversion recovery images. It also demonstrated progressive peripheral enhancement on first-pass perfusion (Video 3) and homogeneous late gadolinium enhancement (Figure 1).

The main differential diagnosis of benign cardiac tumours in infants includes rhabdomyoma, fibroma, and teratoma. Rhabdomyomas are solid tumours that typically show a hyperintense T2-weighted signal, hypoperfusion on first-pass perfusion, and no late enhancement. Fibromas are also solid lesions and usually appear mildly hypointense on T2-weighted images, hypoperfused, and with intense late enhancement. Teratomas, although important to consider in newborns, are usually intrapericardial masses attached to the pulmonary artery and aorta and only rarely intramyocardial. They are heterogeneous tumours with multiloculated cystic and

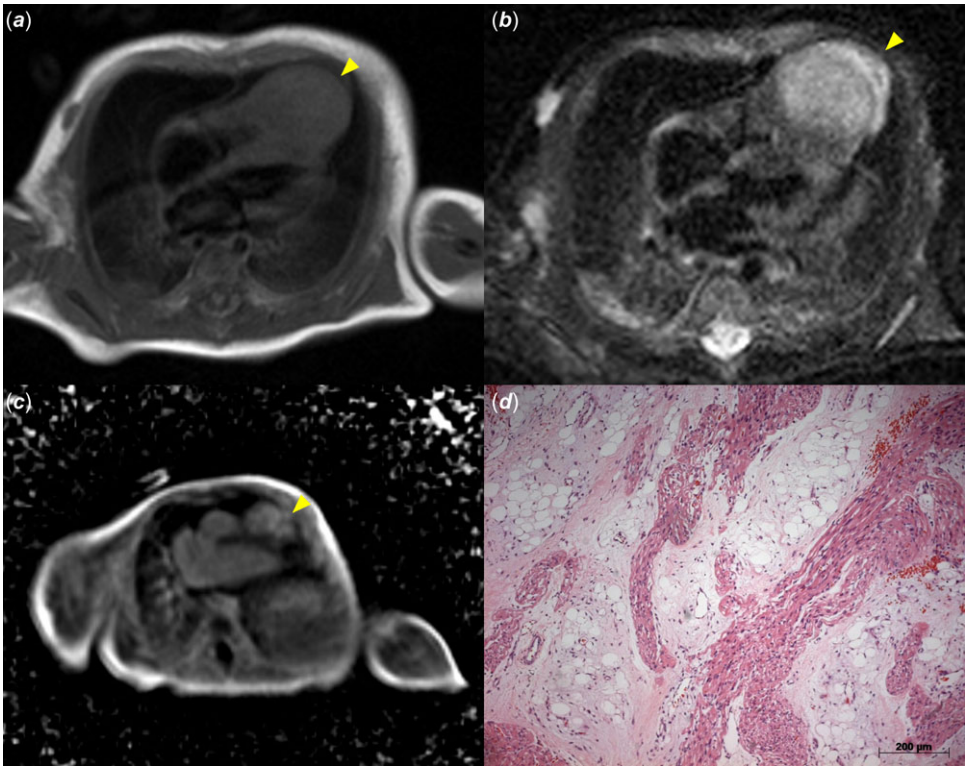


Figure 1. MRI of a newborn with a prenatally diagnosed cardiac mass (***a***–***c***) demonstrating a subepicardial lesion at the right ventricular apex (arrowhead), isointense to the myocardium on T1-weighted double-inversion recovery images (***a***), hyperintense on T2-weighted triple-inversion recovery images (***b***), and showing homogeneous late gadolinium enhancement (***c***). Histopathological analysis revealed bundles of mature cardiomyocytes intermingled with fibroadipose tissue, myxoid areas, and small-vessel proliferation (***d***).

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solid components and are characteristically hypointense on first-pass myocardial perfusion imaging. In this patient, the combination of progressive peripheral enhancement, hyperintense T2-weighted signal, and homogeneous late enhancement was inconsistent with all these entities.

The patient underwent surgical resection, and histopathological analysis revealed disorganised bundles of mature cardiomyocytes intermingled with fibroadipose tissue, myxoid areas, and small-vessel proliferation (D). The cardiomyocytes showed normal morphology, without spider cells or myocyte disarray, which excludes rhabdomyoma and hamartoma of mature cardiomyocytes. The lesion also lacked the dense collagenous stroma characteristic of fibroma. No respiratory or gastrointestinal epithelium, cartilage, bone, neural elements, or other tissues derived from the three germ layers were identified, effectively ruling out teratoma. The presence of multiple mature mesenchymal tissues normally found in the heart supported the diagnosis of cardiac mesenchymal hamartoma, a benign lesion with heterogeneous composition, noninfiltrative behaviour, and favourable postoperative outcome.[1] This tumour is extremely rare and has been predominantly described in the left ventricle. However, the inconsistent terminology in the literature—particularly the overlap between "cardiac hamartoma," "hamartoma of mature cardiomyocytes," and "mesenchymal hamartoma"—precludes a definitive statement regarding the novelty of its right ventricular location.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951126111494.

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Ethical standard. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committees.

Reference

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