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Brief Report

Resection of ventricular rhabdomyomas in infants presenting with cardiac failure

Narendra R. Dereddy,[1] Suvro S. Sett,[2] Usha Krishnan[3]

Departments of [1]Pediatrics, [2]Cardiovascular Surgery, [3]Pediatric Cardiology, Maria Fareri Children's Hospital, Westchester Medical Center, Valhalla, New York, United States of America

Abstract Rhabdomyomas are the most common cardiac tumours in children. They sometimes cause significant obstruction of the ventricular out flow tracts. We report a series of 3 neonates diagnosed antenatally with multiple rhabdomyomas, who developed significant obstruction of the ventricular outflow tracts following birth. They underwent surgical resection in the neonatal period with good outcome. Antenatal diagnosis of obstructive cardiac tumours allows for planning for appropriate early intervention.

Keywords: Cardiac tumours; tuberous sclerosis; ventricular outflow tract obstruction

Rhabdomyomas are the most common cardiac tumours in children, and are usually associated with tuberous sclerosis. They are benign tumours that regress spontaneously, and usually do not require any treatment unless they cause obstruction to the ventricular outflow or inflow tracts, or disturbances of conduction. We report a series of 3 neonates diagnosed antenatally with multiple cardiac rhabdomyomas causing obstruction to the ventricular outflow tracts, which were successfully resected in the neonatal period with excellent results.

Case reports

Baby R was a 4.1 kg, full-term female baby born to a 20 year old mother. Fetal echocardiography during the second trimester had shown multiple right ventricular tumours. Echocardiography after birth showed multiple masses in the right ventricular outflow tract, with obstruction and moderate tricuspid regurgitation (Fig. 1). The baby gradually developed poor feeding with an enlarging liver. Repeated echocardiography at 1 week showed severe obstruction across the right ventricular outflow tract, with a gradient of 80 mm. The tumour was resected through a pulmonary arteriotomy under cardiopulmonary bypass, and the patient made a rapid recovery. Cerebral magnetic resonance imaging showed bilateral frontal cortical tumours, confirming the diagnoses of tuberous sclerosis. Over the subsequent 6 months, her cardiac tumours have regressed, with no residual gradient across the outflow tract.

Baby V was born at term, to a primigravid 25 year old, and had been shown by prenatal echocardiography to have multiple intracardiac masses producing severe obstruction of the left ventricular outflow tract. She also had episodes of fetal supraventricular tachycardia. Due to severe respiratory distress, she was placed on a ventilator soon after birth. The atrial tachycardia was refractory to multiple medications. Echocardiography revealed multiple tumours in the left ventricle, the largest one in the ventricular septum obstructing the outflow tract (Fig. 2), producing a gradient of over 60 mm. In view of possible life-threatening aortic obstruction, she was referred for surgical resection. Under cardiopulmonary bypass, a portion of the septal and valvar tumours obstructing the left ventricular outflow tract was removed. The postoperative period was complicated by recurrent supraventricular tachycardia, which required the addition of flecainide for

Correspondence to: Usha Krishnan, MD, Division of Pediatric Cardiology, 618, Munger Pavilion, New York Medical College, Valhalla, NY 10595, USA. Tel: +1 (914) 493 8372; Fax: +1 (914) 594 4513; E-mail: usha_krishnan@nymc.edu

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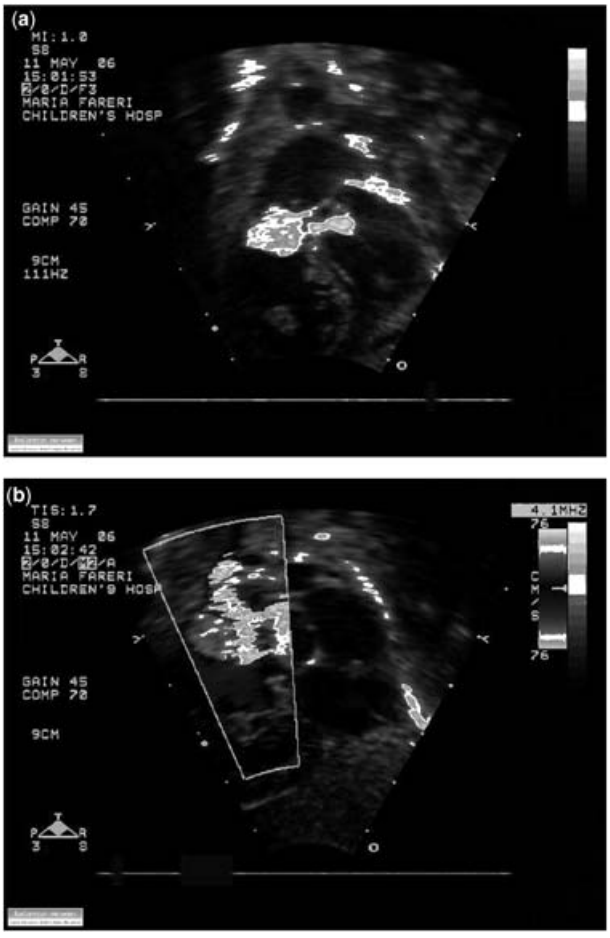

Figure 1.
*Subcostal echocardiogram view (**a**) showing the tumour in the right ventricular outflow tract, with colour flow interrogation (**b**) showing acceleration at the site of obstruction.*

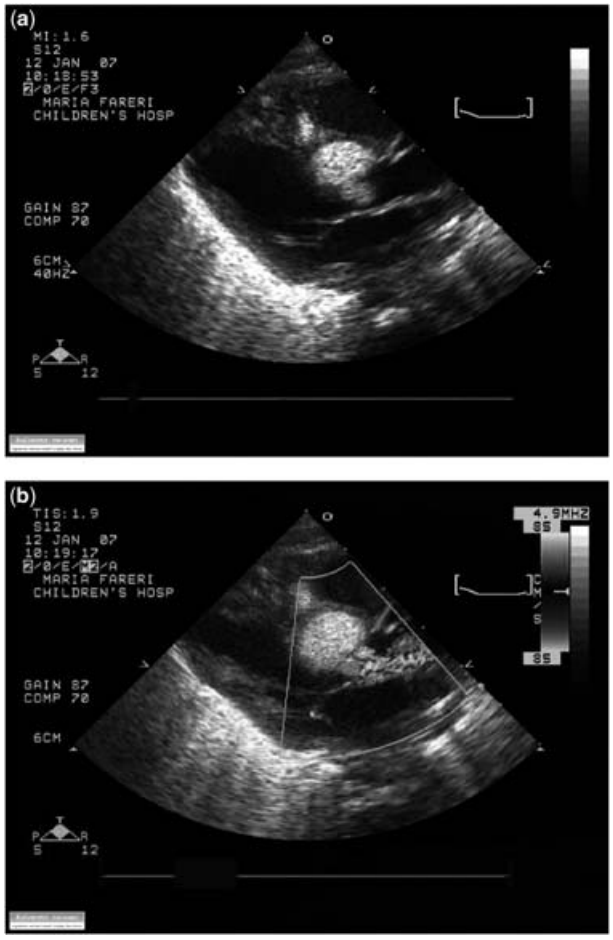

Figure 2.
*Parasternal long-axis echocardiogram view (**a**) showing tumours in the left ventricular outflow tract, with color flow interrogation (**b**) showing acceleration at the site of obstruction.*

treatment. Cerebral magnetic resonance imaging showed bilateral subependymal hamartomas along the lateral ventricles and bilateral frontal cortical tumours consistent with the diagnoses of tuberous sclerosis. After 1 year of follow-up, most of her tumours have regressed, and she is being weaned off anti arrhythmic medication.

Baby S was a full-term baby boy, weighing 4 kg, born with an antenatal diagnosis of multiple cardiac tumours. Postnatal echocardiography showed multiple cardiac tumours, with one of them causing severe obstruction of the left ventricular outflow tract. Due to increasing gradient, the tumour was resected through an aortotomy at 10 days of age. She had no aortic regurgitation post operatively and her tumours were not echocardiographically evident at six months of follow-up. Cerebral magnetic resonance imaging showed subependymal as well as cortical tumours in the left middle frontal gyrus, confirming the diagnoses of tuberous sclerosis.

Discussion

The prevalence rate of cardiac tumours was 0.14% in pregnancies referred for fetal echocardiography, with rhabdomyomas accounting for nine-tenths.[1,2] They are usually multiple, in nine-tenths of cases, and extend into the cardiac cavities in half. They always involve the left ventricle, with right ventricular involvement in four-fifths. They are associated with tuberous sclerosis in up to four-fifths of cases.[3–6] Children may be totally asymptomatic, or may present variously with congestive cardiac failure, low cardiac output due to intracardiac obstruction to flow, arrhythmias of various types, or with sudden death.[7,8] Surgical intervention is indicated in the event of haemodynamic compromise, as was seen in all our 3 cases. Even if multiple tumours are present, only the haemodynamically significant ones need to be resected, as the other tumours regress spontaneously.[9] In all 3 of our patients, surgery was effective

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in providing complete relief of obstruction, with regression of tumours on follow-up. All patients, however, remain on anti-seizure therapy and early intervention for their neurological problems.

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