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Asymptomatic cardiac rhabdomyoma in neonates: is surgery indicated?

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Abstract

Background

Neonatal Tuberosc sclerosis complex may be associated with symptomatic cardiac rhabdomyomas. Cardiac rhabdomyomas are the most common cardiac tumours. The symptoms include haemodynamic instability and life threatening arrhythmias usually requiring early surgical intervention.

Results

We report a case of a 32 week gestation newborn diagnosed with a right ventricular outflow tract mass and subsequently diagnosed with tuberose sclerosis that needed early surgical intervention.

Conclusion

While this case needed early intervention, the need for surgical intervention in otherwise asymptomatic cases is debatable as neonatal cardiac rhabdomyomas can spontaneously regress.

Introduction

Neonatal Tuberose sclerosis complex is associated with symptomatic cardiac rhabdomyomas. Cardiac rhabdomyomas are the most common cardiac tumours. The symptoms include haemodynamic instability and life threatening arrhythmias usually requiring early surgical intervention. The need for surgical intervention in otherwise asymptomatic cases is debatable as neonatal cardiac rhabdomyomas can spontaneously regress over time. We report a case of a 32 week gestation newborn diagnosed with a right ventricular outflow tract mass and subsequently diagnosed with tuberose sclerosis that needed early surgical intervention.

Patient

A 32 week gestation baby born to a primigravida lady with no significant antenatal, maternal medical or family history with a birth weight 1.66kg (9th-25th centile) and head circumference of 26.5cm (0.4th- 2nd centile) was found on routine day 3 examination with a grade 2/6 ejection systolic murmur but otherwise well. Subsequent echocardiogram confirmed the presence of multiple small echogenic masses on the inter-ventricular septum and right ventricle free wall, with the largest lesion measuring 8.2mm x 7.1mm in the right ventricular outflow tract (RVOT) in an otherwise structurally normal heart (figure 1-3).

Figure 1 Top - Parasternal short axis echocardiography view showing 8.2mmx7.1mm mass in the right ventricular outflow tract. Bottom - Four chamber view with small multiple echogenic lesions.

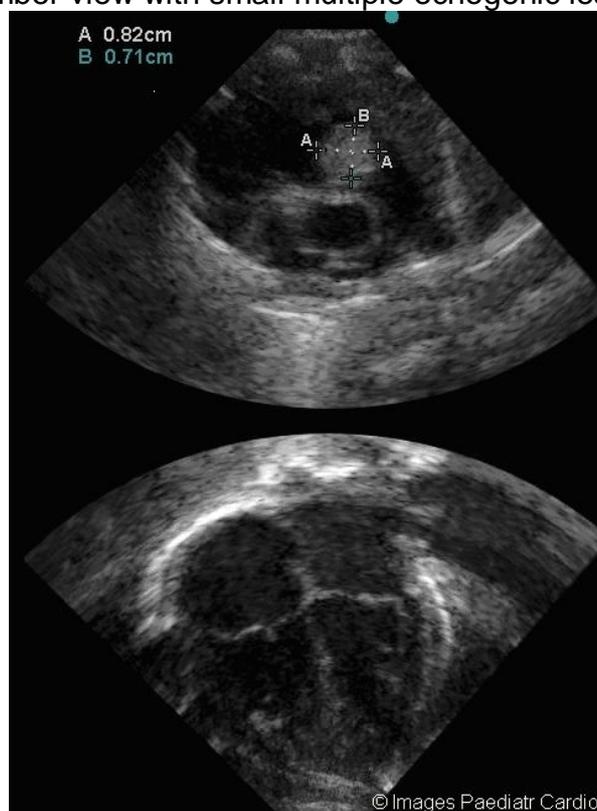


Figure 2 Parasternal long axis echocardiography view of mass in right ventricular outflow tract.

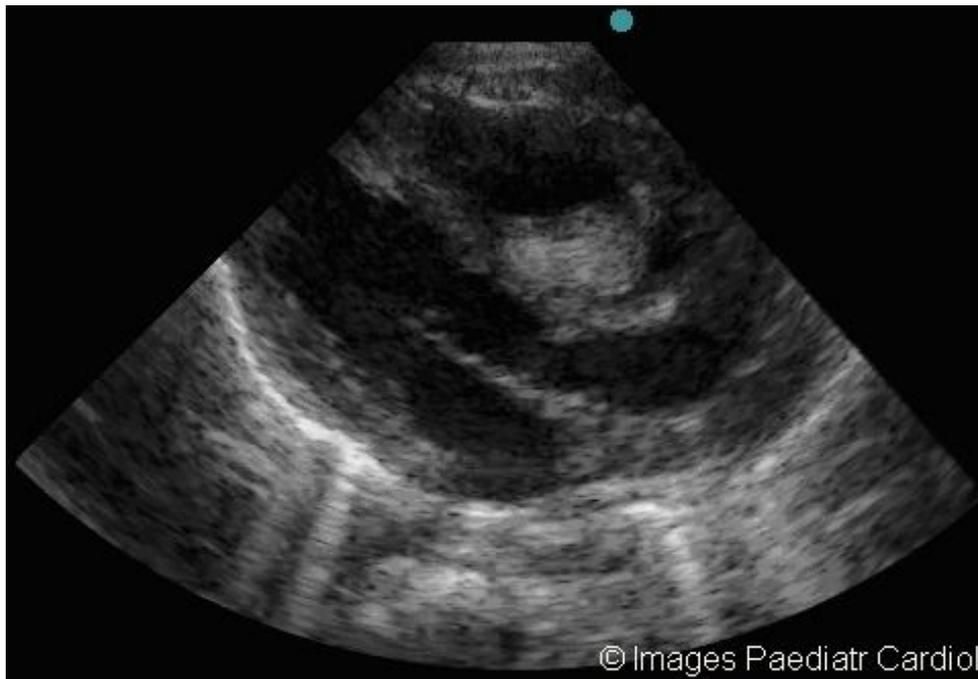
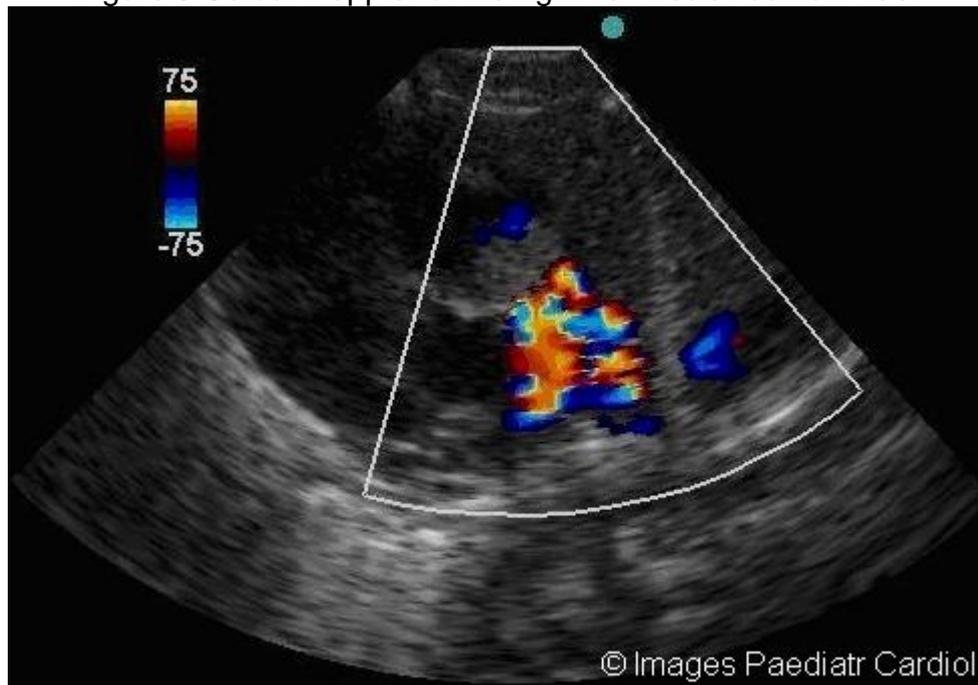
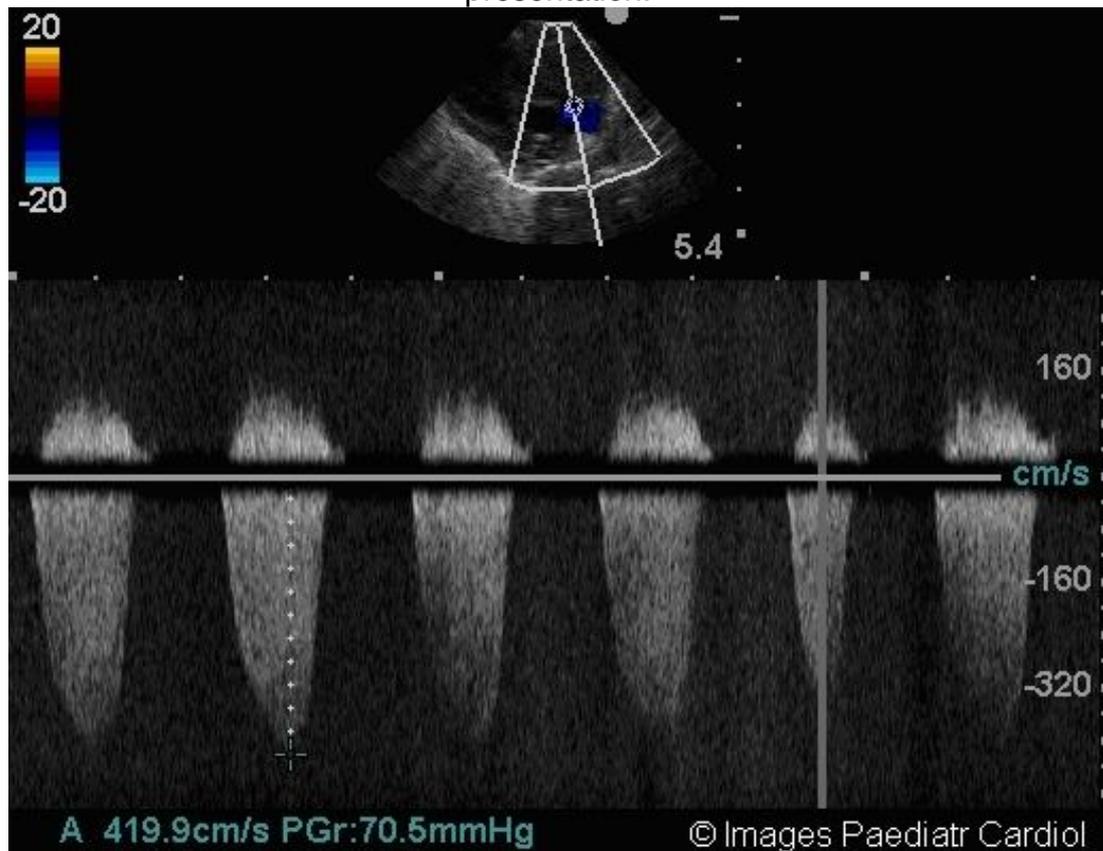


Figure 3 Colour Doppler of the right ventricular outflow tract



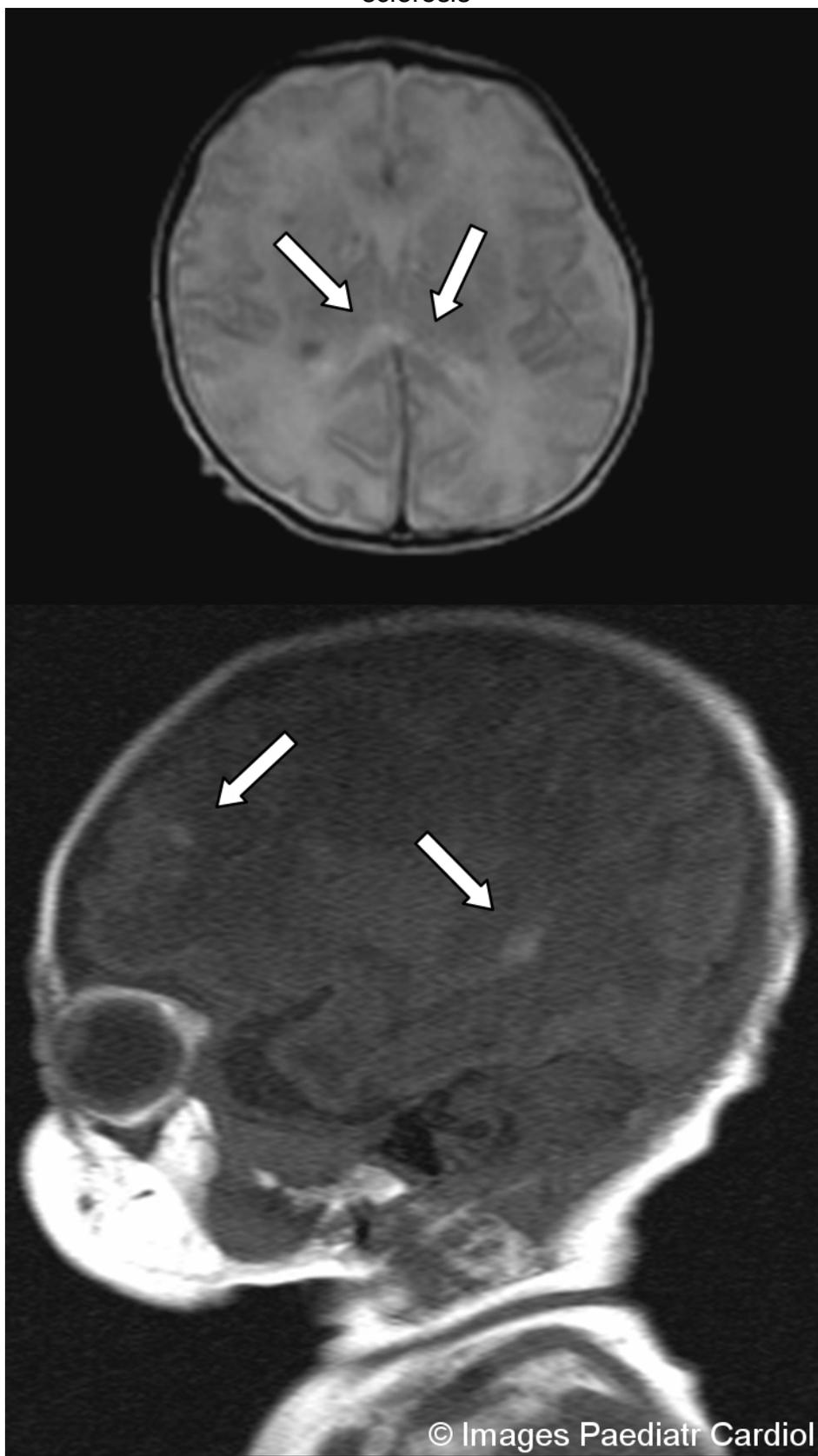
The lesion on the RVOT caused moderate outflow tract obstruction with a Doppler pressure gradient of 70mmHg (figure 4).

Figure 4 Flow acceleration across right ventricular outflow tract at presentation.



Further cardiac evaluation at the paediatric cardiothoracic unit confirmed our findings. Cardiac rhabdomyomas are associated with tuberose sclerosis complex (TSC), and further investigations were performed to evaluate our suspicion in this case. Magnetic resonance imaging (MRI) of the brain showed multiple nodules in the subependymal region and cerebral hemispheres bilaterally (figure 5).

Figure 5 MRI of the brain showing multiple lesions as seen in tuberose sclerosis



Ultrasound imaging of her abdominal organs and Wood's light examination of her skin revealed no lesions. Parental Wood light examination was also normal. These findings are consistent with a clinical diagnosis of TSC. She remained otherwise well and was discharged home from our unit at 36 weeks corrected gestational age. Further cardiac assessment and serial echocardiograms showed a non-regressing mass of 9-11mm in diameter in the RVOT and persistently high Doppler derived peak systolic gradient of 60-98 mmHg. This necessitated surgical intervention at a corrected gestational age of a month to excise the mass in the RVOT and four days later was discharged home after an uncomplicated postoperative period. She continues to thrive with normal developmental milestones at subsequent follow up assessments and serial echocardiograms have shown no reoccurrence on the mass.

Discussion

Cardiac tumours are extremely rare in children (0.027 to 0.17%), and cardiac rhabdomyomas the most common type of cardiac tumours. Cardiac rhabdomyomas are benign fetal hamartomas which are often multiple. They are usually diagnosed in the antenatal period but can present in the neonatal period with haemodynamic compromise although most neonatal cases remain asymptomatic. Cardiac rhabdomyomas are seen in 30-80% of patients with tuberose sclerosis and the natural history is one of spontaneous regression over time.¹ A meta-analysis showed that most rhabdomyomas associated with TSC are non-obstructive.² However they can be obstructive and the majority of obstructive lesions affect the left ventricular outflow tract (LVOT),² while our patient had a less common RVOT lesion. Neonatal cardiac rhabdomyomas causing refractory dysrhythmias or severe haemodynamic compromise are incontrovertible indications for surgery³ but electing to operate in otherwise asymptomatic neonates is debatable. There are few reported cases of right ventricular outflow tract tumours and fewer, possibly under reported cases of asymptomatic rhabdomyomas requiring early surgical intervention. The argument has always been that majority of cardiac rhabdomyomas will spontaneously regress⁴. However some cardiac rhabdomyomas will cause outflow tract obstruction in the absence of life threatening arrhythmias or haemodynamic instability and this may be an indication for early surgery to obviate further haemodynamic compromise. Regression after the neonatal period reduces the risk of reoperation in the long term.⁵

A literature search reveals some reports of early surgical intervention in neonatal cardiac rhabdomyomas. Erdem et al referred their 12 day old baby with a right ventricular outflow tract tumour for early surgery as they could not wait for spontaneous regression because of the significant outflow tract obstruction and the mobility of the mass⁶. Ibrahim et al⁷ were of the opinion that conservative management would be inappropriate in their case and opted for early surgery at 36 hours old as they felt that the morphological appearance of the mass increases the risk of it prolapsing through the aortic valve and potentially causing damage. Freidberg et al reported the case of a 3month old referred for surgery with a large prenatally diagnosed cardiac rhabdomyoma in the RVOT that failed to regress after a period of observation. However peak gradient across the RVOT increased steadily from 40 mmHg to

100 mmHg prompting surgery⁸. Manikoth et al⁹ reported the need for immediate resection on the 5th day of life because of the risk of complete obstruction, embolic event and sudden death. None of the neonates in these reports were haemodynamically compromised or unwell prior to surgery. The common factor appears to be the potential for complication necessitating early surgery and in all cases the surgery was uneventful and patients were discharged home not long after surgery. They were all followed up for suspected tuberose sclerosis. Our patient had serial follow-up echocardiograms, which showed that the mass in the RVOT and close to the pulmonary valves did not regress and continued to cause significant increase in the RVOT pressure gradient. Although she was thriving and showing no signs of haemodynamic compromise after detailed multidisciplinary team discussions and counselling of the parents, early surgical intervention was opted for. She had a good result from her operation and made a smooth and excellent recovery from surgery. She is attaining normal developmental milestones and has shown no other complication from the underlying diagnosis of tuberose sclerosis complex.

In summary, neonatal cardiac rhabdomyomas can spontaneously regress but early surgical intervention in asymptomatic infants may be necessary. Surgery for cardiac tumours carries a mortality of about 5% in children but all the reported cases were uneventful. The choice of management, decision and timing of surgery are largely dependent on the individual case. We recommend that serial echocardiography may help in selecting patients at risk of developing ventricular outflow obstruction and need early surgery and all cases of cardiac rhabdomyomas must be followed up closely for suspected tuberose sclerosis.

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