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Spontaneous Regression of Huge Cardiac Rhabdomyoma in An Infant

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Abstract

Rhabdomyomas are the most common primary cardiac tumors in childhood, and are often associated with tuberous sclerosis. We report a huge rhabdomyoma in an asymptomatic four hour old infant who presented initially with a murmur due to moderate subaortic stenosis. Followup showed regression of the tumour. Although the indications for surgical resection of symptomatic tumors are well established, medical follow-up should be the preferred treatment.

MeSH: Neonate, Rhabdomyoma, Tuberose sclerosis

Introduction

Rhabdomyomas are the most common primary cardiac tumors in childhood.¹⁻³ This neoplasm may be associated with tuberous sclerosis tuberose sclerosis which is an autosomal-dominant disease affecting the brain, skin, kidney, heart and characterised by infantile epilepsy, mental retardation, facial adenoma sebaceum. Although the indication of surgical resection of symptomatic tumors is well established,⁴ medical follow-up is preferred unless critical obstruction or dysrhythmias are present.⁵ We describe a newborn with a huge cardiac rhabdomyoma in the left ventricular outflow tract that initially produced moderate obstruction, but resolved spontaneously.

Case Report

A male neonate aged four hours was admitted to Cerrahpaha Medical Faculty because of a heart murmur detected two hours earlier. Antenatal history and delivery were uneventful and birth weight was 4370g. Physical examination was normal except for a grade 3/6 systolic ejection murmur which was heard maximally at the left sternal border in the third intercostal space. Heart rate was regular at 126/min and blood pressure was 65 mmHg in the right arm.

Echocardiography showed normal chamber dimensions and anatomy, along with a spherical mass originating from the mitral valve, just below the aortic valve, interposed between the ventricular septum and mitral valve anterior leaflet. Multiple additional masses were present within the left and right ventricular cavities (figures 1-3). The location, number and size of each tumor were:

Within right ventricular cavity	
Apical region (figure 1)	8.4 by 5.5 mm
Below septal leaflet of tricuspid valve (figure 2)	4.2 by 8.6 mm
Free parietal ventricular wall	4.2 by 5.6 mm
Within left ventricular cavity	
Obstructing the left ventricular outflow tract	7.2 by 8.2
Interventricular septum (figure 3)	2.2 by 4.2

Figure 1 Mass in right ventricular apex



Figure 2 Mass below septal leaflet of tricuspid valve

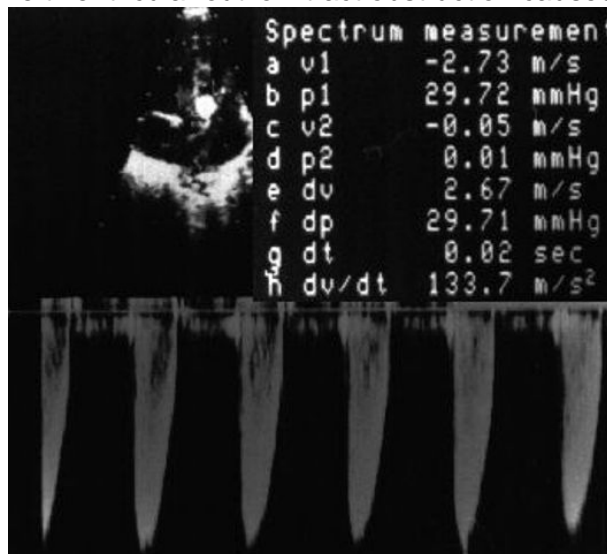


Figure 3 Mass on interventricular septum



Doppler showed turbulence within the aorta with a systolic gradient of 30 mmHg between left ventricle and aorta (figure 4). Although there were no other signs of tuberose sclerosis, the baby was thought to have tuberose sclerosis presenting with heart disease, a common presentation.⁶ Due to the asymptomatic nature of the condition, a conservative approach was taken. Serial echocardiographic studies were undertaken and at one month of age, the subaortic tumor decreased to 7 by 6.8 mm and the outflow gradient fell to 22mmHg.

Figure 4 Left ventricular outflow tract obstruction caused by mass



Neurologic and radiologic features of tuberose sclerosis appeared, with convulsions and subependymal hamartomas at 6 month of age. But cardiologically the patient has remained asymptomatic. Follow-up investigations showed spontaneous regression of the tumors within eight months and the outflow gradient disappeared completely (figure 5). The convulsions were controlled medically and physical and neurologic development appear appropriate for age.

Figure 5 Regression of mass



Discussion

Rhabdomyoma is the most common heart tumor in infancy. A clinicopathologic study showed that such tumors are usually multiple (92%), often intracavitary (50%), and occur more frequently in the left ventricle than in the right ventricle (100 % vs 81 %).² Cardiac rhabdomyomas are frequently associated with tuberose sclerosis,⁶ with a prevalence of 30- 80%.^{1-3,7-12}

The other primary heart tumours at this age (hamartomas, myxomas and fibromas) differ both clinically and ultrasonographically from rhabdomyomas. As most rhabdomyomas are multiple, the diagnosis is generally beyond doubt, and may be made even in the absence of histologic confirmation.¹³ Multiple intracavitary tumours are considered as an important marker of tuberose sclerosis, even in antenatal period.¹⁴ Multiple tumors, or a single tumor plus involvement of other organs (e.g. central nervous system, kidney, skin) that are compatible with the diagnosis of tuberose sclerosis, or a single tumor with positive family history of tuberose sclerosis, is highly suggestive of rhabdomyomas.¹

Cardiac rhabdomyomas may be asymptomatic, or may cause a variety of clinical symptoms depending on their size and location.^{3,15} In the majority of cases, symptoms occur at an early age or even before birth.^{6,14} The spectrum of clinical manifestation ranges from cardiac murmur to sudden death.⁵ The presenting symptom may be arrhythmia, cardiac murmur, complete or variable atrioventricular block, pericardial effusion, cardiomegaly, cardiac failure or sudden death.^{3,6,10,17} The variety of symptoms can be explained on the basis of obstruction of blood flow, myocardial involvement and disturbance of the cardiac rhythm.

The value of surgical resection of symptomatic tumours is well established.⁴ While most rhabdomyomas appear to regress spontaneously, some infants may benefit from surgery for obstructive lesions at an early stage.¹³ With surgery, it is possible to remove obstruction or an arrhythmogenic substrate. Rarely, even heart transplantation may be indicated in patient with severe myocardial involvement.¹⁸ But surgical intervention is neither possible nor indicated in every child.¹⁹ Consequently, a conservative approach is preferable and useful in most cases. Unless critical obstruction or dysrhythmias is present, medical follow-up should be preferred⁵ since these tumours demonstrate benign pathological characteristics and tend to regress over time.^{1,12,13,15,19,20} The chance of spontaneous regression does not depend on the initial size, number or location of tumours.¹⁵ Partial or complete spontaneous regression of rhabdomyomas has been reported in 54% of cases.¹ This regression may take place in a period as short as three weeks.¹

Echocardiography is useful in determining tumour size, number and location. It is also a useful diagnostic method for evaluation of hemodynamic consequence of tumours. Serial echocardiographic studies are both useful and safe in monitoring tumour size, and they provide acceptable follow-up information.²¹

References

1. Beghetti M, Gow RM, Haney I, Mawson J, Williams WG, Freedom RM. Pediatric primary benign cardiac tumors: A 15- year review. *Am Heart J.* 1997;134:1107–1114. [PubMed: 9424072]
2. Fenoglio JJ, McAllister HA, Ferrans VJ. Cardiac rhabdomyoma. A clinicopathologic and electron microscopic study. *Am J Cardiol.* 1976;38:241–251. [PubMed: 952267]
3. Kuehl KS, Perry LW, Chandara R, Scott LP. Left ventricular rhabdomyoma: A rare cause of subaortic stenosis in the newborn infant. *Pediatrics.* 1970;46:464–468. [PubMed: 5454801]
4. Arciniegas E, Hakimi M, Farooki ZQ, Truccone NJ, Green EW. Primary cardiac tumors in children. *J Thorac Cardiovasc Surg.* 1980;79:582–591. [PubMed: 7359937]
5. Farooki ZQ, Ross RD, Paridon SM, Humes RA, Karpawich PP, Pinsky WW. Spontaneous regression of cardiac rhabdomyoma. *Am J Cardiol.* 1991;67:897–899. [PubMed: 2011993]
6. Enbergs A, Borggreffe M, Kurlemann G, Fahrenkamp A, Scheld HH, Jehle J, Breithardt G. Ventricular tachycardia caused by cardiac rhabdomyoma in a young adult with tuberous sclerosis. *Am Heart J.* 1996;132:1263–1265. [PubMed: 8969578]

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7. Bass JL, Breningstall GN, Swaiman KF. Echocardiographic incidence of cardiac rhabdomyoma in tuberous sclerosis. *Am J Cardiol.* 1985;55:1379–1382. [PubMed: 3993573]
8. Gibbs JL. The heart and tuberous sclerosis. An echocardiographic and electrocardiographic study. *Br Heart J.* 1985;54:596–599. [PMCID: PMC481958] [PubMed: 4074592]
9. Mühler EG, Turniski- Harder V, Engelhardt W, von Bernuth G. Cardiac involvement in tuberous sclerosis. *Br Heart J.* 1994;72:584–590. [PMCID: PMC1025649] [PubMed: 7857744]
10. Nir A, Tajik J, Freeman WK, Seward JB, Offord KP, Edwards WD, Mair DD, Gomez MR. Tuberous sclerosis and cardiac rhabdomyoma. *Am J Cardiol.* 1995;76:419–421. [PubMed: 7639176]
11. Shafer RM, Farina M, Alley R, Hansen P, Bishop M. Congenital subaortic stenosis in infancy caused by rhabdomyoma of the left ventricle. *J Thorac Cardiovasc Surg.* 1972;63:157–163. [PubMed: 5008116]
12. Smith HC, Watson GH, Patel RG, Super M. Cardiac rhabdomyomata in tuberous sclerosis: their course and diagnostic value. *Arch Dis Child.* 1989;64:196–200. [PMCID: PMC1791849] [PubMed: 2930225]
13. Webb DW, Thomas RD, Osborne JP. Cardiac rhabdomyomas and their association with tuberous sclerosis. *Arch Dis Child.* 1993;68:367–370. [PMCID: PMC1793857] [PubMed: 8466239]
14. Uzun O, McGawley G, Wharton GA. Multiple cardiac rhabdomyomas: tuberous sclerosis or not ? *Heart.* 1997;77:388. [PMCID: PMC484743] [PubMed: 9155628]
15. Smythe JF, Dyck JD, Smallhorn JF, Freedom RM. Natural history of cardiac rhabdomyoma in infancy and childhood. *Am J Cardiol.* 1990;66:1247–1249. [PubMed: 2239731]
16. Case CL, Gillette PC, Crawford FA. Cardiac rhabdomyomas causing supraventricular and lethal ventricular arrhythmias in an infant. *Am Heart J.* 1991;122:1484–1486. [PubMed: 1951021]
17. Shafer RM, Mintzer J, Alley R, Bishop M. Clinical presentation of rhabdomyoma of the heart in infancy and childhood. *Am J Cardiol.* 1972;30:95–103. [PubMed: 4260837]
18. Demkow M, Sorensen K, Whitehead BF, Rees PG, Sullivan ID, Elliott MJ, de Leval MR. Heart transplantation in an infant with rhabdomyoma. *Pediatr Cardiol.* 1995;16:204–206. [PubMed: 7567670]
19. Foster ED, Spooner EW, Farina MA, Shafer RM, Alley RD. Cardiac rhabdomyoma in the neonate: surgical management. *Ann Thorac Surg.* 1984;37:249–253. [PubMed: 6703807]
20. Alkalay A, Ferry D, Lin B, Fink B, Pomerance J. Spontaneous regression of cardiac rhabdomyoma in tuberous sclerosis. *Clin Pediatr.* 1987;26:532–535.
21. Duncan WJ, Rowe RD, Freedom RM, Izukawa T, Olley PM. Space- occupying lesions of the myocardium: Role of two- dimensional echocardiography in detection of cardiac tumors in children. *Am Heart J.* 1982;104:780–785. [PubMed: 6896956]

