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INTRACARDIAC RHABDOMYOMA IN NEONATES – A CASE SERIES

Neonatology

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ABSTRACT

BACKGROUND: Rhabdomyoma, the most common cardiac neoplasm in neonates. It commonly presents as mechanical obstruction of the outflow tract causing heart failure and arrhythmias. The neoplasm usually undergoes spontaneous resolution. Hence treatment should primarily be symptomatic, while surgical intervention is required in life-threatening situations. Half of these babies develop tuberous sclerosis. There are only few case series available describing its clinical profile especially from our country. Hence, we intended to analyze the same in our institute.

AIM : The aim of our study was to analyze the cases of rhabdomyoma with neonatal presentation in our hospital.

METHODS A retrospective analysis of babies diagnosed as rhabdomyoma was done during the last 15 years (2003 - 2017), based on cardiovascular, radiologic and echocardiographic findings.

RESULTS : Six neonates were diagnosed to have rhabdomyoma in the neonatal period during the last 15 years. Babies who were symptomatic were five (83%) [Two (33%) presented with cardiogenic shock & three (50%) presented with Supraventricular Tachycardia]. One baby was diagnosed based on fetal cardiac mass in antenatal ultrasound. The most common location was the left ventricle and in three babies the tumors were located in multiple chambers. Two (33%) babies had associated tuberous sclerosis. Five babies were managed medically and one required surgical intervention. Four (66%) babies were followed up and were found to be asymptomatic and thriving well.

CONCLUSION: Rhabdomyoma commonest cardiac tumour in neonates is a self limiting condition. Though symptomatic in the newborn period management of low output cardiac failure and arrhythmias leads to better outcome.

KEYWORDS

Rhabdomyoma, Arrhythmias, Tuberous sclerosis.

INTRODUCTION

Rhabdomyoma is the most common heart tumour. Cardiac rhabdomyomas have a natural history of spontaneous regression and are closely associated with tuberous sclerosis complex. This is an autosomal dominant neurocutaneous disorder that can affect every organ of the body, most commonly the brain, Kidney, heart and lungs. Congenital cardiac rhabdomyomas represent a condition of particular interest for the researcher due to spontaneous regression of the tumours that occurs in more than half of cases. Though rhabdomyoma is the commonest cardiac tumour in neonates there are only few case series available describing its clinical profile especially from our country. Hence, we intended to analyze the same in our Institute.

Here, we present the baby with rhabdomyoma presenting with features of congestive cardiac failure in the neonatal period.

CASE SERIES:

- 2 day old male neonate born to a 29 year old primigravida, who had regular antenatal follow up and her antenatal ultrasonography was normal.
- Father was diagnosed to have Tuberous sclerosis in the second decade of life and underwent surgery for intracardiac rhabdomyoma.
- An emergency caesarean section was done in view of fetal distress, baby was depressed at birth requiring 2 cycles of Bag and mask ventilation. APGAR scores were 3 at 1 min and 7 at 5 min.
- Baby was shifted to NICU for post-resuscitation care. Baby had bradycardia and irregularly irregular heart rate with poor peripheral perfusion, was referred to our institution with complaint of arrhythmias and shock.
- **ON EXAMINATION:** Birth weight was 2.8 kg (AGA), a hypomelanotic patch was present over back, tachyarrhythmia's with HR 190/min with features of Congestive cardiac failure.
- **2D-ECHO:** Revealed round, homogenous, hyper-echogenic right ventricular mass, suggestive of rhabdomyoma.
- CT SCAN of brain revealed cortical tubers and diagnosis of tuberous sclerosis was made.
- Baby was managed symptomatically and discharged on day 30 of life. Currently, the baby is 14 months old, asymptomatic, 2D ECHO showed regression in size of rhabdomyoma.

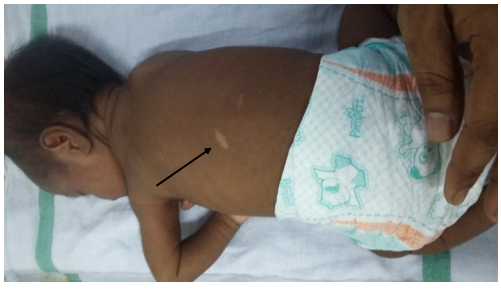


Figure 1: Hypomelanotic Maculae (ashleaf Spots)

Figure 2:adenoma Sebaceum In Father

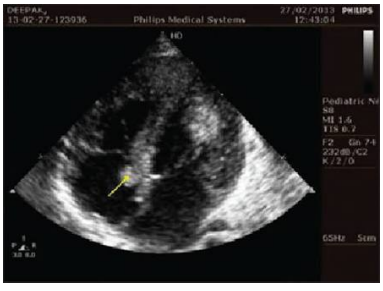


Figure 3: Parasternal Long Axis View Of Mass In Right Ventricular Outflow Tract

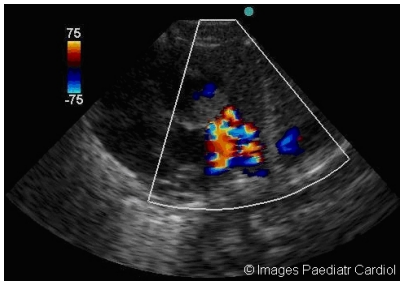


Figure 4: Colour Doppler Of Right Ventricular Outflow Tract

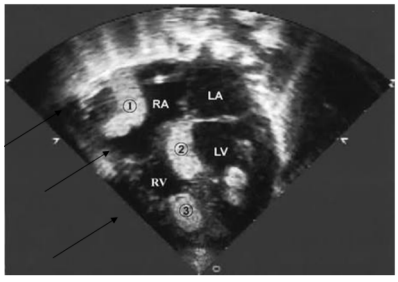


Figure 5:multiple Rhabdomyomas

Supraventricular Tachycardia]. One baby was diagnosed based on fetal cardiac mass in antenatal ultrasound. The most common location was the left ventricle and in three babies the tumors were located in multiple chambers. Two (33%) babies had associated tuberous

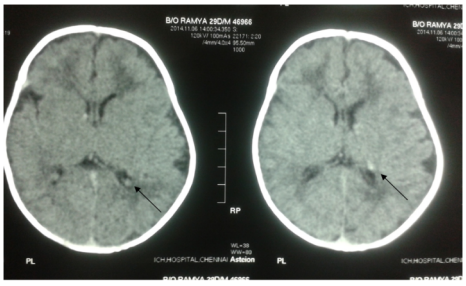


Figure 6: Ct Scan Brain Showing Cortical Tubers

We analyzed the clinical profile of neonates with rhabdomyoma diagnosed in the last 15 years in our hospital.

METHODS

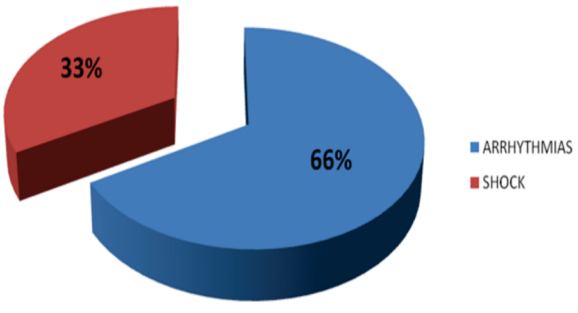
A retrospective analysis of babies with the diagnosis of rhabdomyoma based on cardiovascular, radiologic and echocardiography findings was carried for the period of last 15 years.

RESULTS

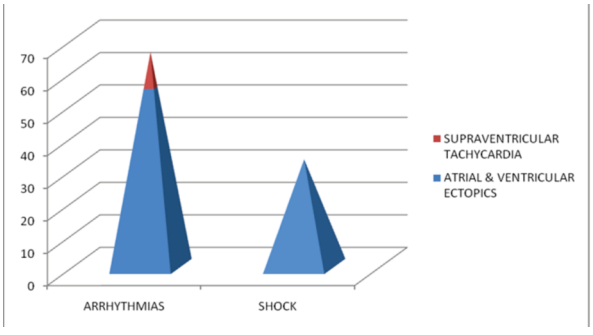
Six babies were diagnosed to have rhabdomyoma during the neonatal period. Babies who were symptomatic were five (83%) [Two (33%) presented with cardiogenic shock & three (50%) presented with sclerosis. Five babies were managed medically and one required surgical intervention. Four (66 %) babies were followed up and were found to be asymptomatic and thriving well.

Table 1: Clinical Presentation – First Cardiac Examination

Case No	Age At Diagnosis	Clinical Signs At First Examination	ECG	ECHO	Cardiac Surgery
1	Day 2	Shock Arrhythmias	Atrial and ventricular ectopic beats	Right ventricle rhabdomyoma	NO
2	Day 2	Arrhythmias	Atrial and ventricular ectopic beats	Multiple rhabdomyomas	NO
3	Day 2	Arrhythmias	RV overload signs	Multiple rhabdomyomas	NO
4	Day 3	Shock	Atrial and ventricular ectopic beats	Left ventricle rhabdomyoma	NO
5	Day 4	Arrhythmias	Atrial and ventricular ectopic beats	Multiple rhabdomyomas	NO
6	Fetal	Antenatal diagnosed cardiac mass	Normal	Multiple rhabdomyomas	NO



Graph 1: Clinical Presentation Of Rhabdomyoma



Graph 2: Cardiac Signs Of Rhabdomyoma

Table 2: General Clinical Presentation – Heart, Brain, Skin, Eye And Kidney Involvement And Family History

Case No	Cardiac Signs	CT Scan	Skin Lesions	Others	Family History
1	Atrial and ventricular ectopic beats **Left ventricle rhabdomyoma**	Cortical tubers	Hypomelanotic maculae	Retinal hamartoma	Positive for Tuberous sclerosis
2	**Arrhythmia** Atrial and ventricular ectopic beats Multiple rhabdomyomas	Cortical tubers	Hypomelanotic maculae	Renal cyst	Negative
3	Arrhythmia RV overload signs Multiple rhabdomyomas	Cortical tubers	Hypomelanotic maculae	Retinal hamartoma	Positive for Tuberous sclerosis
4	Atrial and ventricular ectopic beats **Left ventricle rhabdomyoma**	Subependymal nodules	Facial angiofibroma	Retinal hamartoma	Positive for Tuberous sclerosis
5	**Arrhythmia** Atrial and ventricular ectopic beats Multiple rhabdomyomas	Cortical tubers	Hypomelanotic maculae	No	Negative
6	Multiple rhabdomyomas	Cerebral white matter radial migration lines	Facial angiofibroma, Hypomelanotic maculae	Retinal hamartoma	Negative

DISCUSSION

Rhabdomyomas are the most common cardiac tumours in neonates (45%)[1,2] is an inherited multiorgan disease with incidence of 1 per 5,000 to 10,000 live births can be sporadic and could be associated with tuberous sclerosis complex[3]. Congenital cardiac rhabdomyomas represent a condition of particular interest for the researcher due to spontaneous regression of the tumours that occurs in more than one-half of cases [4]. Partial resolution of the cardiac rhabdomyomas was

reported in 50% of cases and complete regression in 18% and that these tumours grow or appear de novo in 4% of patients with tuberous sclerosis[4]. Surgical resection is not usually considered unless they cause severe intractable arrhythmias, valvular obstruction or congestive heart failure [5].However they are often difficult to be removed completely, because they are usually located deep in the myocardium[6].

Rhabdomyomas appear on ultrasound as round, homogenous, hyperechogenic, intramural or intracavitary masses, sometimes multiple[7], predominantly localized within the ventricles but can be observed in the atria and may lead to obstruction of cardiac valves. They are typically asymptomatic but may also cause atrial or ventricular arrhythmias, sinus node dysfunction and heart block. The involution may be related to the inability of the tumours to divide while the heart chambers grow[8] and this consideration may indicate that some still incompletely identified factors, involved in homeostatic regulation of cardiac biology could lead to regression of the masses. After birth, rhabdomyoma cells lose their ability to divide and regression of the tumour in infancy is an expected outcome, regardless of size of the tumour[9].Complete resolution of more than 80% of the tumours may occur during early childhood [10].

In our case series, rhabdomyomas were mostly present in the ventricles (94%), but also in the right atrium (3%), left atrium (1%) and valves (2%). Most cases were referred due to arrhythmias (66%) or shock (33%) that totally regressed or gradually decreased over time. Only one baby with signs of heart failure was advised for surgical resection but parents refused consent for surgery and was lost for follow-up.

In our case series we noticed involution of rhabdomyomas in all neonates with reduction of size and number, with a decrease of all masses, confirming indirectly the histological observations that these lesions regress. These tumours which originate from the embryonic myocytes represent hamartomas of striated muscular fibres occurring solely in the heart and consists of pathognomonic spider cells with centrally placed cytoplasm containing the nucleus and myofibrils radiating to the cell wall.

FOLLOW UP

4 babies are being followed up and have normal development and none of them had neurological manifestations till date. The cardiac tumours have regressed and the neurological developmental assessment is normal.

CONCLUSION

Rhabdomyoma, the commonest cardiac tumour in neonates is a self limiting condition. If symptomatic in the neonatal period management of low output cardiac failure and arrhythmias leads to better outcome.

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