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Multiple Intracardiac Tumors Causing Obstructive Shock Mimicking Sepsis in a Late Preterm Neonate

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ABSTRACT

Background:
Intracardiac tumors in neonates are rare, with cardiac rhabdomyoma being the most common primary cardiac tumor in this age group. Although many cases are asymptomatic and may regress spontaneously, large or strategically located lesions can result in significant hemodynamic compromise.

Case Presentation:
We report a late preterm male neonate who presented on day three of life with severe respiratory distress, shock, hypoglycaemia, and seizures. Initial evaluation showed presence of thrombocytopenia, metabolic acidosis and acute kidney injury with features suggesting septic shock. The infant was managed by mechanical ventilation, inotropic support, glucose infusion and administration of broad-spectrum antibiotics. Despite intensive care the clinical course was marked by recurrent episodes of desaturation as well as cyanosis. In view of cyanosis not responding to oxygen therapy echocardiography was done which revealed multiple well-circumscribed intracardiac masses involving the left atrium and left ventricle. There was also partial left ventricular inflow obstruction. Features on 2D Echo suggested cardiac rhabdomyoma. The infant subsequently developed refractory sepsis complicated by persistent thrombocytopenia, recurrent bleeding episodes and progressive clinical deterioration. Surgical intervention was deferred due to hemodynamic instability and ongoing sepsis. Despite aggressive supportive management, including blood product transfusions and immunomodulatory therapy, the neonate suffered recurrent cardiac arrest and succumbed on day 21 of life.

Conclusion:
This case highlights a rare as well as fatal presentation of multiple intracardiac tumors causing functional inflow obstruction in a late preterm neonate. It underscores the importance of early echocardiographic evaluation in newborns with unexplained cyanosis accompanied by refractory shock.

Keywords: Cardiac Rhabdomyoma, Echocardiography, Intracardiac Tumor; Myxoma, Neonatal Shock.

INTRODUCTION

Intracardiac tumors in neonates are not that common, with rhabdomyoma being the most common primary cardiac tumor in this age group. Although most cases remain asymptomatic and may regress spontaneously, significant hemodynamic compromise is uncommon.[1] The estimated incidence is at ~1 in 20,000 births and there is a well-known association with tuberous sclerosis, with >50% of all cardiac rhabdomyomas found in patients with later confirmed tuberous sclerosis. The tumors most commonly involve the ventricular myocardium, projecting into the ventricular cavity or moving freely as a pedunculated mass. Rarely, they block circulation within the heart causing heart failure.[2]

Intracardiac tumors often present a diagnostic and therapeutic challenge to treating pediatrics due to their variable clinical manifestations and potential for rapid hemodynamic deterioration despite intensive care . Although many infants remain asymptomatic or regress spontaneously over time, larger or lesions causing obstruction to intracardiac inflow or outflow may lead to significant complications. These complications may include arrhythmias, , hydrops fetalis , inflow or outflow tract obstruction, heart failure or sudden fetal death.[3]

Early recognition through echocardiography is therefore crucial in management of these infants. This is more so in neonates presenting with unexplained respiratory distress, cyanosis or shock. Timely initiation of treatment can potentially reduce disease morbidity, and mammalian target of rapamycin (M-TOR) inhibitors play an effective role in promoting regression of these tumours.[4]

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Although cardiac rhabdomyomas most commonly involve the ventricular myocardium, left atrial involvement leading to functional inflow obstruction in the early neonatal period is uncommon.[5] We report a case of multiple intracardiac masses involving both the left atrium and left ventricle presenting as obstructive shock mimicking septic shock in a late preterm neonate.

CASE REPORT

A late preterm male newborn born at 36 weeks of gestation via lower segment caesarean section (LSCS) presented with severe respiratory distress (Downes score 6), poor perfusion, cold extremities and hypoglycaemia and features suggestive of shock on day 3 of life. The neonate required immediate mechanical ventilation. Furthermore, inotropic support, glucose infusion and antibiotics were also given in view of shock, hypoglycaemia and features s/o sepsis. He developed focal seizures which were managed with intravenous phenobarbitone.

At presentation, the baby showed features of shock with respiratory distress and poor peripheral perfusion. Intermittent episodes of desaturation associated with cyanosis were noted during the hospital course, resolving spontaneously.

Arterial blood gas analysis revealed severe metabolic acidosis (pH 7.264, pCO 5.9, pO247, HCO8.9, oxygen saturation 99%, base excess −24.9). Chest radiograph showed no abnormalities (Figure 1).

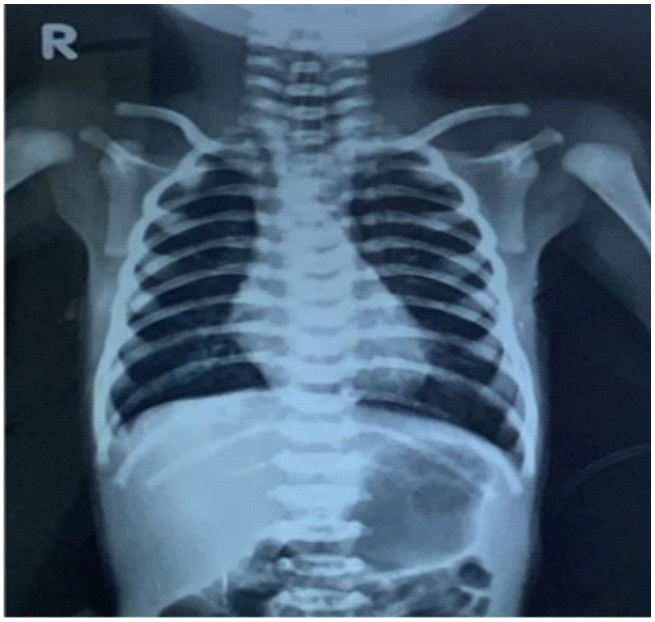

Figure1: Chest radiograph showing normal cardiac silhouette with clear lung fields and no evidence of pulmonary consolidation, pleural effusion, or pneumothorax.

Laboratory investigations demonstrated thrombocytopenia with pre-renal acute kidney injury.

On day 8 of life, repeat complete blood count showed worsening thrombocytopenia with rising C-reactive protein and procalcitonin levels, indicating progressive sepsis. Despite intensive care the clinical course of the neonate was marked by recurrent episodes of desaturation as well as cyanosis. In view of cyanosis not responding to oxygen therapy echocardiography was done which revealed multiple well-circumscribed intracardiac masses involving the left atrium and left ventricle. The largest mass measured 8 × 8 mm in the left atrium, attached to the atrial wall and protruding into the inflow tract. Two additional masses were noted in the papillary muscles as well as left ventricular free wall measuring 6 × 5 mm and 7 × 4 mm respectively.

These masses were seen to be causing left ventricular inflow obstruction. The imaging features were suggestive of rhabdomyoma or myxoma (Figure 2).

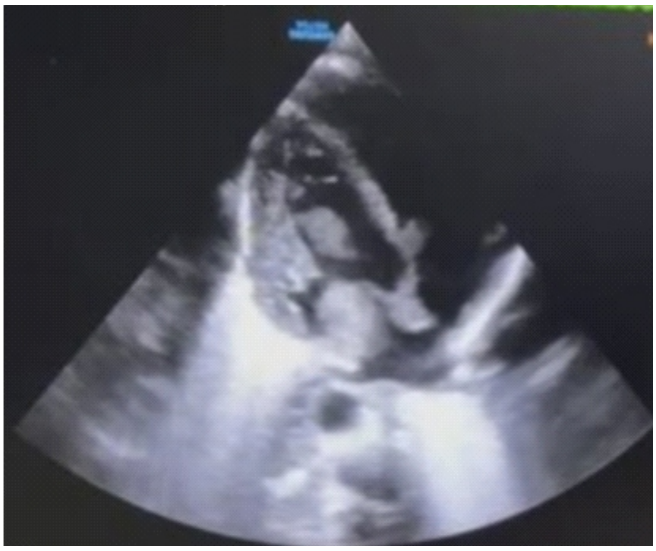

Figure 2: Two-dimensional echocardiography demonstrating multiple well-circumscribed intracardiac masses involving the left atrium and left ventricle. The largest mass is seen arising from the left atrial wall protruding into the inflow tract, with additional lesions in the papillary muscles and left ventricular free wall causing partial inflow obstruction.

Based on echocardiographic findings and recurrent cyanotic episodes, obstructive shock secondary to intracardiac tumors was subsequently suspected.

He was managed with mechanical ventilation, inotropic support, glucose infusion as well as broad-spectrum antibiotics with repeated escalation due to persistent clinical deterioration despite intensive supportive care. Fresh frozen plasma and platelet transfusions were administered for thrombocytopenia and bleeding episodes. Pediatric cardiology and cardiothoracic surgery opinions were sought who recommended surgical removal of the masses once the neonate becomes hemodynamically stable.

On day 9 of life the baby suffered cardiac arrest and was successfully resuscitated. Ongoing sepsis with worsening thrombocytopenia required repeated platelet transfusions. Intravenous immunoglobulin therapy was initiated on day 19 due to presence of refractory sepsis not responding to antibiotic therapy.

Despite aggressive supportive care, antimicrobial escalation, and blood product and immunoglobulin transfusions the infant continued to deteriorate. On day 21 of life, he developed massive oro-nasal hemorrhage requiring fresh frozen plasma and packed red cell transfusions followed by sudden cardiac arrest. Resuscitation efforts were unsuccessful and the patient succumbed.

DISCUSSION

Primary cardiac tumors in neonates are rare with an estimated incidence of less than 0.3% in autopsy series. Among these, rhabdomyomas are the most frequently encountered followed by fibromas and myxomas . These tumors are often detected incidentally however in some cases they may cause significant hemodynamic compromise due to inflow or outflow obstruction, arrhythmias or thrombo-embolic events.[6]

In this case multiple well-circumscribed intracardiac masses involving both the left atrium and left ventricle were causing functional inflow obstruction. This inflow obstruction manifested as episodes of recurrent desaturation as well as cyanosis.

Although the infant initially presented with features of septic shock, intermittent hypoxic episodes and echocardiographic findings indicated a cardiac etiology.[7] This emphasizes the importance of considering possibility of structural cardiac diseases in neonates with refractory shock or unexplained desaturation even when primary diagnosis is something else. Reports of cardiac rhabdomyomas presenting predominantly as refractory septic shock and episodes of intermittent cyanosis are very scarce and therefor a high index of suspicion from treating neonatologists for possibility of these lesions is important for early diagnosis and timely management of these cases.[8]

In this case multiplicity of lesions and ventricular involvement favoured rhabdomyoma as the most likely diagnosis. However, histopathological confirmation was not possible due to rapid clinical deterioration. Evaluation for tuberous sclerosis complex including neuroimaging and genetic testing could not be performed because of the infant's critical condition. Although surgical intervention is indicated in cases with significant obstruction or life-threatening complications, hemodynamic instability and ongoing sepsis excluded possibility of operative intervention. Moreover, the coexistence of severe sepsis and intracardiac tumors significantly worsened prognosis in this case.[9]

Persistent thrombocytopenia, recurrent bleeding episodes, rising inflammatory markers and poor response to antimicrobial therapy indicated refractory septic shock. In addition, features suggestive of consumptive coagulopathy were also present in this case. Presence of Obstructive cardiac physiology because of mechanical obstruction caused by intracardiac tumors further compounded systemic hypoperfusion thereby accelerated multiorgan dysfunction. Despite aggressive supportive care (including mechanical ventilation, Inotropic support, blood product transfusions and intravenous immunoglobulin) the infant progressed to irreversible shock and succumbed. While most cardiac rhabdomyomas regress spontaneously and known to have favourable outcome early fatal presentation can be secondary to combined hemodynamic obstruction as well as systemic infection.[10]

This case underscores the need for early echocardiographic evaluation in neonates who present with disproportionate or refractory shock. In these cases, prompt identification of structural cardiac disease may influence management and outcomes. Lack of histopathological confirmation remains a limitation of this report.

CONCLUSION

This case demonstrates a rare presentation of an intracardiac mass causing inflow obstruction and clinically mimicking sepsis in a late preterm neonate. These tumors caused mechanical inflow obstruction thereby causing recurrent cyanotic episodes and refractory shock. This case also underscores the importance of early echocardiographic evaluation in neonates with refractory shock and unexplained recurrent cyanotic spells. A high index of suspicion on the part of treating neonatologist may help in early diagnosis and timely management. Early recognition of structural cardiac diseases including intracardiac tumors may influence management strategies including possibility of timely surgical interventions.

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