"Amber in chamber" an enigmatous right atrial mass in a neonate

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ABSTRACT

The incidence of fungal infection is increasing worldwide. Although fungal infection is common in adults, few cases have been reported in the neonatal population. We report a case of the preterm neonate of 34 weeks who developed respiratory distress on 2nd day and was initiated on mechanical ventilation. Treatment was instituted for sepsis, but the patient continued to deteriorate. Two-dimensional echocardiography revealed a large right atrial mass, which eventually turned out to be fungal ball. Intense surgical and medical management led to a speedy recovery of the patient. We stress on the early use of echocardiography in atypical presentation of neonatal septicaemia along with routine investigations to help in early recognition of source of infection. This can be of great value in initiating definitive management and improving survival rate in such patients.

Key words: Fungal endocarditis, neonatal septicemia, right atrium, two-dimensional echocardiography

INTRODUCTION

In today’s world the menace of fungal infections is rising day by day. Fungal infections are associated with high mortality and morbidity. Candida is the most common fungal pathogen in human beings. According to the 1-day point prevalence Extended Prevalence of Infection in Intensive Care II study conducted over 75 countries in 2007, Candida was the third most common pathogen with an infection rate of 17% after Staphylococcus aureus and Pseudomonas.[¹] Owing to the low incidence but high mortality of fungal endocarditis in the neonatal population, early diagnosis and treatment is utmost important. We report a rare case of neonatal sepsis with right atrial mass which eventually turned out to be fungal ball.

CASE REPORT

A 34 weeks old preterm neonate weighing 2.1 kg was delivered by emergency lower segment cesarean section following abruptio-placenta. Next day, the patient developed respiratory distress for which mechanical ventilation was initiated and was initiated on mechanical ventilation. Treatment was instituted for sepsis, but the patient continued to deteriorate. Two-dimensional echocardiography revealed a large right atrial mass, which eventually turned out to be fungal ball. The mechanical ventilation was initiated with FiO₂ 60%, respiratory rate 30/min; tidal volume 20 ml, positive end-expiratory pressure-3 cm H₂O, inspiratory to expiratory ratio (I:E 1:1.5) maintaining SpO₂ 90-95%. After few hours of mechanical ventilation the patient started deteriorating, two-dimensional (2D) echocardiography was done which showed a large right atrial mass obstructing right ventricular inflow. Finally, the decision for intra-cardiac excision of the
mass was taken as it was obstructing the right ventricular inflow and compromising outflow from the right ventricle to the pulmonary artery. Patient was shifted to the operation theater with all hemodynamic monitors in situ. Induction was achieved by the inhalational technique using sevoflurane, i.v. fentanyl 8 µg and rocuronium 2 mg. The sternum was opened through standard median sternotomy, cardiopulmonary bypass instituted after heparinization and right atrium exposed. A grayish white mass of 3 cm × 3 cm, which was almost completely closing the tricuspid valve and destroying septal leaflet was identified as in Figure 1. The mass was excised by sacrificing septal leaflet of the tricuspid valve, and the defect was repaired, keeping foramen ovale patent for spontaneous closure.

After completion of the surgery, the patient was weaned off from cardiopulmonary bypass with inotropic support of milrinone (0.5 mcg/kg/min), dobutamine (10 mcg/kg/min), adrenaline (0.1 mcg/kg/min), noradrenaline (0.03 mcg/kg/min) and nitro-glycerine (0.5 mcg/kg/min), to support right ventricle in view of decreasing right ventricular afterload to avoid right ventricular failure. Immediate postoperative transesophageal echocardiography showed moderate to severe tricuspid regurgitation with no residual vegetation. The patient was shifted to Intensive Care Unit (ICU) and the specimen was sent for microbiological culture and histopathological examination. Potassium hydroxide preparation showed budding yeast cells and culture showed colonies of Candida albicans as shown in Figure 2. There was a mixed infection of Aspergillus and Candida, and hence antifungal liposomal amphotericin B according to National Institutes of Health dosage regime was started in a dose of 3 mg/kg/day [Table 1].

On 4th postoperative day, 2D echocardiography showed mild to moderate tricuspid regurgitation. The patient condition gradually improved and was extubated on 5th postoperative day. On 8th postoperative day all inotropes were tapered off with patient vitals stable and maintaining oxygen saturation of 96% on room air. The child gradually became more active, playful and was accepting breastfeeds thereafter discharged on postoperative day 20.

**DISCUSSION**

Owing to the low incidence but high mortality (75-90%) of fungal endocarditis in the neonatal population, early diagnosis and treatment is of utmost important. Major cause of fungal septicemia in neonatal ICU is Candida and may affect 1.6-12.9% of particularly very low birth weight patient, that is, birth weight <1500 g. In last decade, the incidence of fungal endocarditis is on the rise secondary to more use of central venous line placement, broad spectrum antibiotics, and neonatal surgical interventions. In the current study, the neonate had low immunity (was preterm)

**Table 1: Recommended anti-fungal dosing in neonates**

| Agent                  | Dose                  | Side effect                                      |
|------------------------|-----------------------|--------------------------------------------------|
| Amphotericin B deoxycholate | 1 mg/kg/day           | Infusion-related toxicity: Anaphylactic reactions, Nephrotoxicity, Hypokalemia |
| Liposomal amphotericin B | 3 mg/kg/day to 5 mg/kg/day | Fewer infusion related reactions and nephrotoxicity |
| Caspofungin            | 25 mg/m²/day          | Hepatotoxicity, fever, headache, rash, gastrointestinal symptoms, anemia |
| Micafungin             | 10 mg/kg/day          | Hepatotoxic, gastrointestinal symptoms: Nausea and vomiting |
| Anidulafungin          | 1.5 mg/kg/day         | Nausea, vomiting, rashes, thrombophlebitis        |

**Figure 1:** Right atrial mass with right ventricular inflow obstruction

**Figure 2:** Budding yeast like cells suggestive of Candida species seen in Gram’s-stained smear
along with it he was on broad spectrum antibiotics and on mechanical ventilation. These factors all combined would have to have led to fungal endocarditis in this neonate. Among all the reported cases, two-thirds of these cases of fungal endocarditis are due to Candida.[7] Blood culture is often negative in such patients, and less than one-half of candidal endocarditis cases yield positive blood cultures. The complete blood count may reveal leukocytosis, thrombocytopenia, raised erythrocyte sedimentation rates and C-reactive protein.

In neonates, symptoms are often nonspecific and include bradycardia, apnea, hypothermia, feeding intolerance, increased ventilatory support, poor perfusion and evidence of septic emboli and rarely a new or changing heart murmur. Sometimes cardiac involvement, without other symptoms or signs of infection, may be the only clinically apparent feature.

Two-dimensional echocardiography, may be helpful in early diagnosis and management as it provides real time imaging of the entire right atrium, interatrial septum, inferior venae cavae, superior venae cavae and tricuspid orifice. There are very few reported cases regarding the use of echocardiography in neonatal fungal endocarditis thereof resulting surgical removal of the mass and patient survival. Rubinstein et al. reported a case of a premature infant with a central hyperalimentation catheter in the right atrium. The baby had a large thrombus on the catheter with the help of echocardiography it was diagnosed and thereafter successfully eradicated with urokinase-induced thrombolysis.[8] Riggs et al. initiated echocardiogram studies in all patients with candidiasis and were able to detect right atrial masses in 6 cases, 5 of those were preterm.[9] All patients had some degree of perinatal asphyxia, needed mechanical ventilation, received broad spectrum antibiotics and parenteral nutrition with lipid emulsions. Therefore, echocardiography may play a vital role in early search of nidus. Approximately, one-fourth of neonates and children with systemic candidal disease have a demonstrable cardiac lesion. Hence, we stress on the early use of echocardiography in neonates with atypical presentation of septicemia along with routine investigations to help in early recognition of source of infection. This can be of great value in initiating definitive management and improving survival rate in such a patient.

Aggressive antifungal therapy is always necessary but may not prove sufficient to completely alleviate the infection, so removal of the infected nidus becomes central to the management. We wish to emphasize that wherever specific symptoms are present, such as on-going infection (not responding adequately to medical treatment), embolic phenomena, and cardiac decompensation, delaying surgical intervention is not advantageous. Although a small number of patients have survived with medical therapy alone, most survivors have required both medical and surgical treatment.

CONCLUSION

Fungal infections are infrequent but has a fatal outcome, particularly in neonates. In cases where neonatal sepsis is not responding to conventional therapy, there should not be any delay in echocardiographic evaluation to find out the source of infection. This should be followed by medical treatment or surgical removal of the nidus, which can be lifesaving.

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How to cite this article: Kumar S, Sharma JP, Saigal S, Dhurwe R. “Amber in chamber” an enigmatous right atrial mass in a neonate. Saudi J Anaesth 2015;9:477-9.
Source of Support: Nil, Conflict of Interest: None declared.