SHORT COMMUNICATION

Unusual cause of neonatal cyanosis

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Abstract We present a case of a full-term female neonate who presented at 6 h of age with severe cyanosis and was partially responsive to oxygen supplementation. An echocardiogram showed an isolated congenital severe tricuspid valve insufficiency due to rupture of the papillary muscle of the anterior tricuspid valve leaflet. Magnesium sulfate was infused to lower the pulmonary resistance and thus enhancing the antegrade pulmonary blood flow. Ductal patency was secured by prostaglandin infusion thus providing an additional pulmonary blood flow through the ductus arteriosus.

The above measures were adequate to stabilize the patient with no further deterioration or the need for other supportive measures such as Nitric Oxide therapy or extracorporeal membrane oxygenation (ECMO). Therefore, early diagnosis and adequate measures to improve the pulmonary blood flow are mandatory, important pre-operative measures in the management of these patients.

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1. Introduction

Tricuspid valve regurgitation (TR) due to chordal rupture is an extremely rare differential diagnosis of neonatal cyanosis (Riede et al., 2010).

2. Case report

A full-term female neonate was delivered at 38 weeks gestation by normal spontaneous vertex delivery and weighed 3900 g at birth. Patient’s APGAR scores were 9, 10 at 1 and 5 min, respectively. Meconium staining was noticed during neonatal resuscitation. Pregnancy was uneventful and normal ultrasound was normal at 22 weeks of gestation. The mother did not notice any decrease in the fetal movements during the last trimester. She did not take drugs during the last trimester of pregnancy even including prostaglandin synthetase inhibitors and she did not have a history suggestive of systemic lupus erythematosus.

Six hours after birth the neonatology team noticed severe cyanosis and abdominal distention. The initial diagnosis was meconium aspiration syndrome. However, patient cyanosis did not improve by 100% oxygen supplementation.

Prompt cardiology consultation was made to rule out congenital heart disease. Cardiovascular examination revealed a
distressed, cyanosed, nondysmorphic newborn. The heart rate was 140/min, blood pressure was 66/34 mm Hg and the respiratory rate was 46/min. The initial oxygen saturation was 70% which increased to 80% with 100% oxygen inhalation via head-box. The capillary refill time was about 3 s and peripheral pulses were equally felt. The liver edge was palpated 4 cm below the right costal margin and the liver itself was abnormally pulsating. Sacral edema and ascites were present. The precordium was hyperactive. Cardiac auscultation revealed normal first and second sounds, no gallop rhythm and a harsh pan-systolic murmur of grade 3/6 was heard at the left lower sternal border.

Chest X-ray revealed oligemic lung fields and there was cardiomegaly. Twelve leads standard ECG was performed and revealed a sinus rhythm at a rate of 160/min, left axis deviation, prominent P wave suggestive of P-pulmonale, no evidence of left or right ventricular hypertrophy. There were no ST segment changes, and the corrected QT interval was within normal limits. Cardiac echocardiography using Vivid 7 ultrasound system (GE, Horten, Norway) revealed normal segmental anatomy. The right atrium was significantly dilated. A right to left shunt was detected through a stretched patent foramen ovale. The right ventricle was hypertrophied. The diagnosis of rupture of the tricuspid valve (TV) papillary muscle was made after identification of flail antero-superior leaflet of the TV with a thickened echogenic tip and the absence of a connection between the anterior papillary muscle and the flail leaflet of TV (Fig. 1). The tip of the anterior papillary muscle appears echo bright. Severe TV regurgitation was present (Fig. 2) and the tricuspid valve regurgitation pressure gradient was 55 mm Hg. The antegrade pulmonary flow was reduced in the color Doppler mode, however, no detectable pressure gradient across the pulmonary valve was elicited. The ductus arteriosus was closed and no area of calcification was present in the usual ductal location. The right atria was dilated and right to left shunt was observed through a stretched patent foramen ovale (PFO). There were no vegetations or thrombi. CK and CK MB enzymes were within normal limits. No laboratory evidence of maternal lupus was found.

The patient was promptly stabilized by inotropes, prostaglandin E1, magnesium sulfate and Lasix therapy. 100% oxygen (7 l/min) inhalation by headbox was sufficient to maintain her oxygen saturation at 90%. The above measures were adequate to stabilize the patient with no further deterioration or need for other supportive measures such as Nitric Oxide therapy or ECMO.

Repeated examination at 24 h after birth revealed improved patient cyanosis which allowed the gradual withdrawal of oxygen therapy and inotropic support. Repeated echocardiography at the age of 2 weeks confirmed the diagnosis. The shunt across the patent foramen ovale (PFO) became mostly left to right. Sufficient antegrade and minimal retrograde pulmonary blood flow was seen. Surgical intervention will be planned electively in the near future.

3. Discussion

Neonatal tricuspid valve insufficiency frequently complicates other forms of congenital heart disease (Kobza et al., 2004; Kanter et al., 2004). In contrast there are few case reports in the literatures regarding neonatal critical tricuspid regurgitation due to ruptured papillary muscle or chordae (Anagnostopoulos et al., 2007 Apr; Katogi et al., 1998).

Most of the reported cases as well as the present case presented with congestive heart failure and cyanosis (Lim et al., 2004). The tricuspid regurgitation in these cases may be severe enough to preclude antegrade pulmonary flow, with a resultant massive right to left atrial shunt (Lim et al., 2004).

The differential diagnosis of papillary muscle rupture can be broadly divided into ischemic and non-ischemic etiologies (Fleming et al., 2008).

Non-ischemic causes include trauma and infective endocarditis (Fleming et al., 2008). Both can be ruled out in our case since the delivery was uneventful and there was no clinical or laboratory findings consistent with the infection. In addition there was no evidence of any vegetation in the immediate post-delivery echocardiogram.

Myocardial ischemia may result from antepartum or peripartum asphyxia (Riede et al., 2010), viral infections (Marton et al., 2002; Lazda et al., 2000), Rhesus isoimmunisation (Marton et al., 2002) and maternal auto-immune disease.
The papillary muscles are particularly vulnerable to ischemia because they lie at the distal extremes of coronary circulation and require a greater oxygen supply than subepicardial myocardium (De Busk and Harrison, 1969).

In the present case, the delivery was carried out smoothly, patient’s Apgar score was acceptable and patient’s cardiac enzymes were within the normal range thus eliminating the possibility of perinatal ischemia.

Although the cause of papillary rupture is unclear in our case, however, one might speculate on transient ischemia during the pregnancy. The echo bright flail appearance of anterior tricuspid valve leaflet (Fig. 1) suggests in utero insult rather than an acute perinatal insult (Sachdeva et al., 2007). Antenatal ductal closure is of interest beyond its importance in limiting treatment options. If closure occurred suddenly, a tremendous increase in right ventricular afterload would result during severe fetal stress, and the combined hemodynamic and metabolic insult might provoke ischemic rupture of the tricuspid papillary muscle (Sachdeva et al., 2007). However, there was no history of late gestational intake of prostaglandin synthetase inhibitors (PSI) in our patient to explain the premature ductal closure. Chronic in utero ductal closure is unlikely in the present case because no area of calcification was present in the usual ductal location.

The marked cyanosis observed immediately after birth is due to marked right to left shunt at the atrial level due to high pulmonary resistance. MgSO₄ infusion was given in the present case to lower the pulmonary resistance and thus reducing right to left shunt at the atrial level as well as to enhance the antegrade pulmonary blood flow. Ductal patency was secured by Prostaglandin E₁ infusion thus providing an additional retrograde pulmonary blood flow through the ductus arteriosus. Such prompt management was successful in improving patient’s cyanosis. This signifies the importance of the time factor in initiation of these simple measures to avoid the expense of Nitric Oxide and complication of ECMO.

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References


