Case Report

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When Four Lesions Collide: Neonatal Shone's Complex Unfolded

Fatimazahra Guissi*, Kaoutar Danaoui, Fatimaezzahra Tahiri, Hajar Akki, Kaoutar Ettoini, Mouna Zouine and Abdallah Oulmaati

Neonatal Intensive Care Unit, Mohammed VI University Hospital Center of Tangier, Faculty of Medicine and Pharmacy, Abdelmalek Essaadi University, Tangier, Morocco

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***Corresponding author:** Fatimazahra Guissi, Neonatal Intensive Care Unit, Mohammed VI University Hospital Center of Tangier, Faculty of Medicine and Pharmacy, Abdelmalek Essaadi University, Tangier, Morocco, E-mail: dr.guissifatimazahra@gmail.com

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ABSTRACT

Background: Shone's complex is a rare congenital heart disease characterized by multiple levels of left-sided obstruction, including a supravalvular mitral ring, parachute mitral valve, subaortic stenosis and coarctation of the aorta. The complete form is exceptionally uncommon and may present during the neonatal period with severe heart failure or cardiogenic shock.

Case Presentation: We present a case of full-term male neonate admitted to the Neonatal Intensive Care Unit (NICU) at Mohammed VI University Hospital in Tangier for respiratory distress. Echocardiography confirmed the diagnosis of complete Shone's complex. The newborn received multidisciplinary management including prostaglandin infusion, respiratory support and diuretics. Despite intensive care, the infant developed cardiogenic shock and died on day 17th of life. Parental consent was obtained.

Conclusion: This case underscores the importance of maintaining a high index of suspicion for rare congenital heart diseases in critically ill neonates, as well as the value of rapid echocardiographic diagnosis and timely intervention in complex cardiac anomalies such as Shone's complex.

Keywords: Shone's complex, Cardiogenic shock, Congenital heart disease, Neonate, Echocardiography

Abbreviations: NICU: Neonatal Intensive Care Unit; CT: Computed Tomography; CPAP: Continuous Positive Airway Pressure; POCUS: Point-of-Care Ultrasound; CTA: Computed Tomography Angiography

1. Introduction

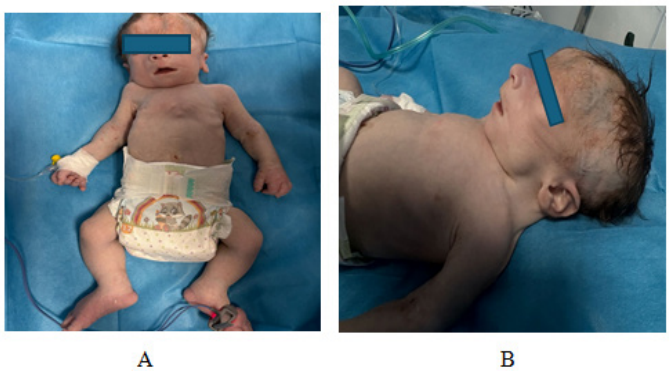
Shone's complex is a rare congenital cardiac anomaly characterized by multiple levels of left-sided obstruction, classically involving a supravalvular mitral ring, parachute mitral valve, subaortic stenosis and coarctation of the aorta[1]. The complete form, comprising all four lesions, is exceedingly uncommon and often presents in early infancy with signs of heart failure or cardiogenic shock[2]. Early diagnosis remains challenging due to overlapping clinical features with more common neonatal conditions, including respiratory distress syndrome and sepsis.

Advances in echocardiography and Point-of-Care Ultrasound (POCUS) have significantly improved early recognition of such complex lesions[3]. We report a case of neonatal cardiogenic shock caused by complete Shone's complex, emphasizing the diagnostic value of bedside echocardiography in the acute care setting.

2. Case Presentation

Here we present a case of male newborn was admitted to our Neonatal Intensive Care Unit (NICU) on day 1 of life for respiratory distress. Antenatal ultrasound performed at 32 weeks of gestation had revealed a single umbilical artery and hypoplasia of the aortic isthmus. The pregnancy was otherwise uneventful. Delivery was vaginal at 39 weeks of gestation, with a birth weight of 3,200 g and Apgar scores of 10 at 1 and 5 minutes.

At 6 hours of life, the infant developed progressive tachypnea (respiratory rate: 70 breaths/min) with hypoxemia (SpO_2: 82% on room air) poorly responsive to supplemental oxygen therapy. Physical examination revealed axial hypotonia, dysmorphic features including low-set ears, short broad neck and hypertelorism. Cardiopulmonary auscultation was unremarkable, with no murmur detected initially. Peripheral pulses were symmetrically palpable but weak. Blood pressure was 65/40 mmHg in the right upper limb. Initial laboratory investigations revealed metabolic acidosis (pH: 7.28, lactate: 4.2 mmol/L), with normal complete blood count and C-reactive protein levels.



A B

Figure 1 A and B: Clinical presentation of a newborn with Shone's complex.

2.1. Echocardiographic Findings

Transthoracic echocardiography performed within the first 12 hours of life revealed features consistent with complete Shone's complex: Supravalvular mitral ring with a mean gradient of 8 mmHg parachute mitral valve with a single papillary muscle and moderate stenosis, bicuspid aortic valve with minimal regurgitation, hypoplastic aortic annulus, suspected aortic coarctation at the isthmus level, patent ductus arteriosus with bidirectional shunting, severe pulmonary arterial hypertension (estimated systolic pulmonary artery pressure: 65 mmHg), left ventricular function preserved (ejection fraction: 58%).

2.2. Computed Tomography Angiography (CTA)

Computed Tomography Angiography (CTA) confirmed severe aortic coarctation at the isthmus with a minimal diameter of 2.8 mm, persistent ductus arteriosus, cardiomegaly (cardiothoracic ratio: 0.68) and signs of pulmonary hypertension. An additional finding was a single left pulmonary vein draining into the left atrium.

2.2.1. Management: The newborn was managed with Continuous Positive Airway Pressure (CPAP) ventilation, prostaglandin E1 infusion (0.05 µg/kg/min) to maintain ductal patency and intravenous furosemide (1 mg/kg twice daily). Fluid restriction was implemented.

2.2.2. Clinical course: On day 8th, the infant developed fever with elevated inflammatory markers, consistent with nosocomial infection. Blood cultures grew coagulase-negative *Staphylococcus*. Broad-spectrum antibiotic therapy with vancomycin and gentamicin was initiated.

Despite appropriate antimicrobial therapy and continued supportive care, the infant's hemodynamic status progressively deteriorated. On day 15th, he developed overt cardiogenic shock with hypotension (mean arterial pressure: 28 mmHg), oliguria and worsening metabolic acidosis. Emergency intubation and mechanical ventilation were performed and inotropic support with dobutamine (10 µg/kg/min) was initiated. The infant's condition continued to decline despite maximal medical therapy, the patient died on day 17th of life.

3. Discussion

Shone's complex is a rare congenital heart defect first described by John Shone and colleagues in 1963[1]. Its prevalence among all congenital heart diseases is estimated at approximately 0.6% to 0.7%[4]. Orphanet classifies Shone syndrome as an extremely rare disease, with a prevalence of less than 1 per 1,000,000[5]. Despite its rarity, the condition may be underdiagnosed, particularly when patients present later in life or with incomplete forms.

Classically, Shone's syndrome is defined by the presence of four left heart obstructive lesions: supravalvular mitral membrane, parachute mitral valve, subaortic stenosis and coarctation of the aorta[1]. These anomalies can present in a complete form (all four lesions present) or an incomplete form (two or three lesions), which directly influences the prognosis and surgical approach[6]. However, recent studies have reported a wider spectrum of associated anomalies, including valvular aortic stenosis, ventricular septal defects, mitral annular hypoplasia, patent ductus arteriosus and other left ventricular abnormalities[4,7,8]. As a result, some authors describe up to six to eight associated lesions, highlighting the heterogeneity of the condition and the complexity of its anatomical presentation in neonates[9].

In our case, the patient presented with supravalvular mitral membrane, parachute mitral valve, bicuspid aortic valve with

hypoplastic annulus, coarctation of the aorta and patent ductus arteriosus, consistent with the extended variant of Shone's syndrome. This emphasizes the need for comprehensive echocardiographic assessment and individualized management strategies.

In neonates with Shone's syndrome, the clinical presentation is typically severe, including respiratory distress, congestive heart failure and poor feeding[10]. Some individuals may remain asymptomatic, with symptoms developing only in adolescence or adulthood[11]. Qatza, et al.[10] reported a 4th week-old infant presenting with tachypnea (40 breaths/min), tachycardia (150 beats/min), hypoxia (SpO□: 80%) and blood pressure of 90/55 mmHg in the upper limbs, while lower extremity measurements were unobtainable. Jordan, et al.[3] described a 6th week-old infant who developed cardiogenic shock with severe respiratory distress, similar to our case.

Echocardiography remains the cornerstone for early detection and detailed evaluation of Shone's complex[12]. Recent neonatal case reports demonstrate how early transthoracic echocardiography can identify multi-level obstructive lesions, guide initial management and aid surgical planning. Prompt echocardiographic assessment in neonates with unexplained respiratory distress or cardiovascular instability improves the likelihood of diagnosing complex congenital cardiac anomalies[10].

Prenatal echocardiography plays a crucial role in the early detection of left-sided obstructive lesions, including those seen in Shone's complex. Fetal echocardiography can identify structural abnormalities such as mitral valve obstruction, aortic coarctation and ventricular hypoplasia, allowing early diagnosis and perinatal management planning[13]. Prenatal diagnosis is associated with improved neonatal outcomes, as it enables delivery in specialized centers equipped for prompt postnatal intervention. In our case, antenatal detection of aortic isthmus hypoplasia facilitated early postnatal evaluation, although the complete diagnosis was established only after birth.

The prognosis of complete Shone's complex in the neonatal period remains challenging. Although surgical techniques have advanced considerably, infants with multiple left-sided obstructive lesions often require early intervention and may still experience high morbidity and mortality[14]. Surgical options include staged repair with initial coarctation repair followed by subsequent valve interventions or comprehensive primary repair in selected cases[6]. The decision depends on the severity of each lesion, left ventricular size and function and overall clinical status.

Ahmed, et al.[14] conducted a systematic review of surgical outcomes in patients with Shone's complex and reported that outcomes depend heavily on the severity of obstruction, the number of lesions and the timing of intervention. Neonates with complete forms and severe obstruction at multiple levels carry the highest risk. Additionally, nosocomial infections, as observed in our patient, can significantly complicate the clinical course and delay surgical intervention.

Although Shone's complex is rare, studies encompassing broader age ranges indicate a wide clinical spectrum. Older patients with incomplete or late-diagnosed variants may present with chronic complications such as arrhythmias, heart failure or aortic dissection, emphasizing that the spectrum and outcomes vary significantly with age at presentation and completeness of lesion involvement[11,15].

4. Conclusion

Shone's complex is a rare and severe congenital heart disease requiring early recognition and thorough cardiac assessment. This case emphasizes the importance of prompt multidisciplinary management in neonates presenting with unexplained respiratory distress or cardiovascular instability. Echocardiography is essential for early diagnosis and surgical planning. Prognosis depends on the extent of cardiac anomalies, associated comorbidities and timing of intervention. Reporting such cases improves understanding of this complex condition and supports evidence-based clinical decision-making.

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