

Case Report

Double aortic arch presenting as acute upper airway obstruction in a Nigerian newborn: A case report.

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Accepted 39 November, 2010

Aortic arch abnormalities manifesting at birth are rare. Most reported cases present with symptoms later in life with features of respiratory obstruction presenting much earlier than those of oesophageal obstruction. Patients with significant airway compression may die as a result of respiratory compromise, but such events are said to be extremely rare. We report the case of a baby with double aortic arch who manifested with features of severe upper airway obstruction from birth and succumbed to the effect of severe hypoxia before any surgical intervention could be effected. This report is intended to create awareness of the condition in Nigeria and highlight the importance of early recognition of newborns with double aortic arch so that appropriate measures to overcome airway obstruction could be initiated to avert significant hypoxia-related complications.

Keyword: Aortic arch, Cardiac anomaly, Nigerian Newborn

INTRODUCTION

Aortic arch anomalies are rare (Chan et al., 2003) congenital vascular conditions that may present with apparent life-threatening events. Double aortic arch is the most common type of congenital vascular ring anomalies (Chan et al., 2003). The reported incidence of aortic vascular rings was 0.3-0.6% in one series (Cordovilla et al., 1994). Little is known regarding the exact cause of double aortic arch. The association of the condition with chromosome 22q11 deletion implies that a genetic component is likely in certain cases (Momma et al., 1999).

Double aortic arch may occur in isolation or in association with other congenital heart defects (Chan et al., 2003; Cordovilla et al., 1994; Momma et al., 1999; Bakker et al., 1999). The clinical features are primarily dependent on the degree of airways and/or oesophageal compression. Severe distress can occur in newborns at birth but most cases do not manifest significant symptoms until later in life when tracheomalacia or lung complications might have set in (Bakker et al., 1999; Tuo et al., 2009). Although the disease is characterized by long term morbidity, early surgical intervention is associated with excellent outcome (Cordovilla et al., 1994).

There is paucity of data on double aortic arch in Nigeria. Various studies on congenital heart diseases in Nigeria ((Asani et al., 2005; Bode-Thomas et al., 2003; Jaiyesimi and Antia, 1991; Olowu, 1995) have not cited a case of double aortic arch. This might be the first report of the condition in Nigeria. This report is intended to create awareness of the existence of the disease in our locality and highlight the importance of early recognition of newborns with double aortic arch so that appropriate measures to overcome airway obstruction could be initiated before hypoxic injuries ensue.

CASE REPORT

A 7-day old baby-girl was referred to our unit with the history of noisy breathing, chest in-drawing, darkish discoloration of lips and extremities since birth, cough and fever of a day duration. The Pregnancy was supervised at a Primary Healthcare Centre. Mother had severe dyspepsia in the third trimester warranting the use of antacid suspension (milk magnesium trisilicate). No history of use of any other drugs. No other maternal illness during pregnancy. The baby was delivered at 40 weeks gestation at home by breech extraction. She did not cry immediately after birth until after about an hour of mechanical stimulation. She was then noticed to have

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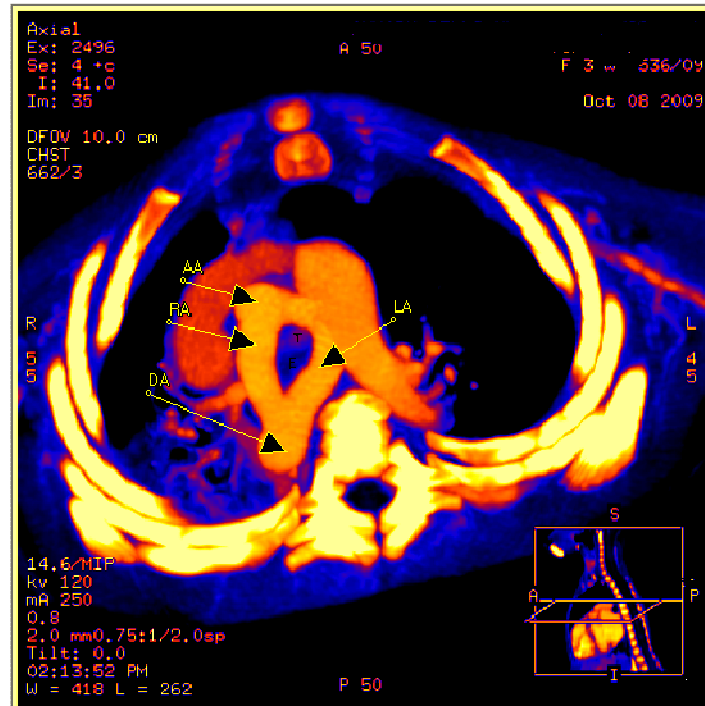


Figure 1. CT angiography showing right (RA) aortic arch and left (LA) aortic arch completely encircling the trachea (T) and esophagus (E). The ascending aorta (AA) and descending aorta (DA) are also displayed

very weak cry, noisy breathing and darkish discoloration of the lips that tended to be worsened when baby cried. Breathing was laboured with chest in drawing. There was no body swelling and no abnormality noticed in any part of the body.

Breastfeeding was initiated at the 12th hour of life. Baby sucked with difficulty and had intermittent apnoeic attacks during breastfeeding but no choking or drooling of saliva and no vomiting or abdominal distention. The baby was the 4th child of the mother, a 25-year old housewife, in a consanguineous marriage relationship with 45 years old man.

At presentation, the baby was moderately dehydrated and in severe respiratory distress: tracheal tugging, marked intercostal and subcostal recessions and was centrally cyanosed. Her respiratory rate was 60 per minute. She was also having intermittent brassy cough and loud inspiratory stridor. The breath sound was vesicular but of reduced intensity. There were occasional coarse crepitations on both lung fields. The cardiovascular findings were unremarkable except for tachycardia of 160 per minute. Her blood pressure was 60/40mmhg. Examination of other systems was normal apart from tender hepatomegaly of 7cm. Her anthropometry was normal (weight = 3.1 kg, length = 51 cm, head circumference = 35 cm).

The initial assessment was severe bronchopneumonia with congestive cardiac failure in an infant who had had

some degree of perinatal asphyxia. She was investigated and managed accordingly with supplemental O₂, antibiotics, frusemide, digoxin and caloric supply. She showed a slight improvement but stridor and cyanosis persisted. SpO₂ was 40-60% at room air but improved to 99% on supplemental O₂. The diagnosis of congenital upper airway obstruction was then considered: laryngomalacia, subglottic stenosis/haemangioma and vascular ring being uppermost. Radiological evaluation of the upper airway and vascular tree was then carried out.

Her laboratory results revealed haematocrit of 36%, total leucocyte count of $6.4 \times 10^9/l$ with 72% neutrophils, 26% lymphocytes, 2% monocytes and a platelet count of $205 \times 10^9/l$. Results of infection screening were negative. Serum biochemistry showed urea of 17.0 mmol/l, sodium of 127 mmol/l, potassium of 4.2 mmol/l, chloride of 86 mmol/l and bicarbonate of 24 mmol/l. The serum calcium was 2.36 mmol/l, phosphorus was 1.68 mmol/l, total protein was 66g/l and albumin was 39g/l. Echocardiographic study revealed normal ventriculoatrial and ventriculoarterial concordance but with an ostium secundum defect measuring 0.36 cm. Angiography showed right and left aortic arches completely encircling the trachea and oesophagus and the descending aorta was on the right (Figure 1). The right aortic arch was larger than the left and gave off the right common carotid and subclavian arteries. Lateral chest X-ray showed narrowing of the trachea air column just above the carina.

The infant was being prepared for emergency surgery when she suddenly had a prolonged apnoeic attack that failed to respond to resuscitation and was certified dead.

DISCUSSION

Tracheal compression by vascular structures is uncommon in newborns and may be masked by non-specific respiratory symptoms. Therefore a high index of suspicion is necessary for diagnosis. Symptoms of respiratory obstruction at birth or soon after should alert the Physician to the possibility of vascular ring obstruction (Chan et al., 2003).

Double aortic arch results from failure of regression of both the right and left fourth branchial arches, resulting in right and left aortic arches, respectively. Normally, absorption of the right (posterior) arch takes place between the right subclavian artery and its junction with the descending aorta. The remnant of the right arch becomes the right innominate artery and leaves a left (anterior) arch in normal development, freeing the trachea and oesophagus. Failure of resorption of the right arch produces a vascular ring that encircles and compresses the oesophagus and trachea leading to severe respiratory and feeding difficulties (Chan et al., 2003).

Double aortic arch is commonly an isolated anomaly but may occur in association with other congenital heart defects (Momma et al., 1999). Of the commonly reported cardiac anomalies, ventricular septal defects ranked first followed by atrial septal defects, patent ductus arteriosus, tetralogy of Fallot and transposition of great arteries (Lee, 2007). Our patient had an ostium secundum atrial septal defect.

Infants with double aortic arch commonly present with stridor (100%), cough (75%), dyspnoea (75%), reflex apnoea (60%), respiratory tract infections (56%) and feeding difficulties (25%) (Gormley et al., 1999). The index case had all these symptoms and, in addition, was in cardiac failure probably due to the effects of severe hypoxia on the myocardium as well as the associated bronchopneumonia. It is therefore recommended to evaluate every newborn or child with stridor, wheezing, and dysphagia for the presence of a complete vascular ring early before complications set in (Lee, 2007).

Tracheal compression may cause significant morbidity and mortality from severe hypoxia, an effect that could be obviated with continuous positive airway pressure ventilation (CPAP) while awaiting surgery, (Chan et al., 2003) most facilities in developing countries, however, lack the wherewithal for CPAP.

Definitive diagnosis of double aortic arch is usually achieved with radiological studies. Although there is no pathognomonic feature of double aortic arch on plain chest radiographs, chest X-ray may show a deviation or compression of the trachea, or contour of a right arch,

especially in the scenario of an infant presenting with respiratory and gastroesophageal symptoms (Lee, 2007). The lateral X-ray of the chest of our patient showed narrowing of the tracheal air column just above the carina, thus corroborating the report of other authors (Lee, 2007). Barium oesophagography has been advocated as a valuable investigative tool for patients with suspected vascular rings and it was once considered the single most important method of study in evaluating patients with vascular rings (Van Son et al., 2004). But the advent of newer imaging techniques like computed tomography (CT) and Magnetic resonance (MRI) has rendered barium oesophagography less appealing as the former is safer and provides a more accurate diagnosis (Van Son et al., 2004). Contrast computerized tomography done in our patient confirmed the diagnosis of double aortic arch thus rendering the need for barium oesophagoscopy unnecessary.

In patients with double aortic arch presenting with symptoms of airway or oesophageal compression, early surgical division of the smaller aortic arch is recommended. Postoperative outcome was excellent in a series by Cordovilla *et al* (Cordovilla et al., 1994) as far back 1994. Our case, however, succumbed to the effects of hypoxia before surgical intervention could be performed. This is to buttress the lack of high-tech ventilatory support facilities and emergency preparedness in most resource-limited settings like our hospital.

Although our case was an early neonatal mortality, two out of the six patients in Lee's series (Lee, 2007) tolerated respiratory and feeding problems from birth to 14 and 15.5 years of age, respectively, thus signifying that sufferer of this disease have ample chance of surviving beyond the neonatal period even in the absence of surgical intervention. The reason for the early neonatal death in our case is not certain but may not be unconnected with the associated cardiac failure, hyponatraemia and severe azotaemia.

In conclusion, our report emphasizes the consideration of vascular abnormalities induced respiratory distress in neonates; even as they encompass a relatively rare group of disorders capable of considerable morbidity and mortality if not diagnosed and managed in time.

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