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Double Aortic Arch Presented with Neonatal Stridor†

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CASE HISTORY

This one day old female neonate was brought to Sung-Ae Hospital because of noisy respiration and mild dyspnea on February 17, 1994. She was born to a primiparous mother via c-section because of cephalopelvic disproportion at 9:25 AM, February 17, 1994. The gestation age was 38^{+5} weeks and birth weight was 3. 4kg.

According to the report from the private Ob & Gyn Clinic where she was born, a mild dyspnea and grunting had been present since immediately after birth. There was no premature rupture of the membrane. Otherwise his general condition was fair. However, the dyspnea and grunting showed no improvement. She was transferred to Sung Ae Hospital approximately 10 hours after delivery.

At the time of admission the baby's general activity and crying were good. She was not cyanotic. Respiration was noisy, but the grunting was not definite. Physical examination showed no external anomalies. Head and neck were unremarkable. The chest showed symmetric expansion. A mild subcostal retraction was present. The breathing sound was clear but inspiratory stridor was present. No murmur was audible. The abdomen was slightly distended and soft. No palpable mass was noted. The bowel sounds were normal. The back and extremities were unremarkable. Neurological examination revealed all reflexes to be active and symmetrical.

Chest X-ray showed cardiomegaly and normal pulmonary vasculature. Focal air trapping was noted in the right middle and lower lung fields. Laryngoscopy revealed that the epiglottis that appeared to be sucked in during inspiration.

After admission the baby developed cyanosis. Inspiratory stridor and expiratory wheezing became apparent. She was put in a 40% O_2 hood. Arterial blood gas analysis showed pH 956, PCO_2 88.2 mmHg, pO_2 117.4 mmHg, HCO_3 19.6 mmol/l CO_2 content 22.2 mmol/l, base excess -15.4 mmol/l and O_2 saturation 95.1%.

The cyanosis did not improve with the oxygen hood despite O_2 being increased to 60%. Subsequent ABGA showed persistent CO_2 retention. She was placed on ventilator care (RR 50/min, PIP 12, PEEP 2, FiO_2 0.4). Blood pressure was 90/50 mmHg.

Three hours after admission her inspiratory stridor continued to be aggravated, and chest X-ray showed decreased aeration and left lung atelectasis. Cyanosis and bradycardia became apparent. ABGA showed pH 6.809, PCO_2 160.5 mmHg, PO_2 33.9 mmHg, BE -14.7 mmol/l and

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HCO_3 25.5 mmol/l. The ventilator setting was changed to FiO_2 0.5, RR 65, PEEP 5. However, her general condition showed no improvement. Chest X-ray showed increased heart size(Fig 1). Routine laboratory data including hemogram, bilirubin, C-reactive protein and electrolytes all remained within normal range. The patient expired at 8:30 AM February 18, 1994.

DISCUSSION

Dr. B. Kim, In summary, this term newborn infant with AGA showed clinical manifestations of progressive respiratory distress without perinatal problems immediately after birth.

In general, many different categories of diseases and conditions should be considered in differential diagnosis of respiratory distress in newborn infants (Table 1). However, in this case some categories of primary lung diseases, extrinsic compression of the lung, and nonpulmonary causes can easily be excluded by the clinical history, physical findings and laboratory data.

Table 1. Differential diagnosis of respiratory distress in neonate

I. Airway obstruction
- A. Nasal or nasopharyngeal
  - 1. Choanal atresia
- B. Oral cavity
  - 1. Macroglossia
  - 2. Micrognathia
  - 3. Pierre Robin syndrome
- C. Neck
  - 1. Congenital goiter
  - 2. Cystic hygroma
- D. Larynx
  - 1. Laryngomalacia(congenital laryngeal stridor)
  - 2. Vocal cord paralysis
  - 3. Congenital laryngeal stenosis
  - 4. Laryngeal atresia
  - 5. Congenital subglottic stenosis
  - 6. Subglottic hemangioma
  - 7. Laryngo-tracheo-esophageal cleft
- E. Trachea
  - 1) Intraluminal and intramural
    - 1. Tracheomalacia
    - 2. Tracheal stenosis
    - 3. Tracheal agenesis
    - 4. Tracheoesophageal fistula with or without esophageal aresia
  - 2) Extramural
    - 1. Vascular ring or sling
    - 2. Tumor or cyst
- F. Bronchus
  - 1) Intramural and intraluminal
    - 1. Congenital bronchial stenosis
  - 2) Extramural
    - 1. Bronchogenic cyst

II. Primary lung disease
- 1. Transient tachypnea of the newborn
- 2. Aspiratory syndrome (meconium, amniotic fluid)
- 3. Respiratory distress syndrome
- 4. Pneumonia
- 5. Pulmonary hemorrhage
- 6. Primary atelectasis
- 7. Pulmonary lymphangiectasia
- 8. Pulmonary hypoplasia or aplasia
- 9. Congenital cystic adenomatoid malformation
- 10. CLD

III. Extrinsic compression of the lung
- 1. Extrapulmonary air leak
- 2. Congenital lobar emphysema
- 3. Pleural effusion, chylothorax
- 4. Congenital diaphragmatic hernia
- 5. Intrathoracic cyst or tumor
- 6. Thoracic dystrophy

IV. Nonpulmonary causes
- 1. Congestive heart failure
- 2. Metabolic cause(acidosis, hypothermia, hypoglycemia)
- 3. Persistent pulmonary hypertension of newborn
- 4. Birth asphyxia
- 5. CNS or neuromuscular disease

The cause of rapidly progressive respiratory distress can be considered as obstruction, and the site of obstruction can be considered to be the trachea on the grounds of the characteristic symptoms and physical findings of respiratory noises including inspiratory stri-

dor and expiratory wheezing in spite of endotracheal intubation, absence of facial abnormality or neck mass, symmetric expansion of the chest and clear breathing sound, and normal laryngoscopic finding except that of the epiglottis which appeared to be sucked in during inspiration.

Here I can narrow down the causes of rapidly progressive respiratory distress, characterized by initial ventilatory failure and later respiratory failure, into intrinsic and extrinsic tracheal obstruction (Table 2).

Table 2. Causes of inspiratory stridor and expiratory wheezing in neonates

A. Intrinsic obstruction
  1. Tracheal stenosis
  2. Tracheomalacia
B. Extrinsic obstruction
  1. Vascular ring or sling
    1) double aortic arch
    2) right aortic arch with left ductus or ligamentum arteriosum or left aortic arch with right ductus or ligamentum arteriosum
    3) anomalous innominate or left carotid artery
    4) aberrant right subclavian artery
    5) right aortic arch with aberrant left subclavian artery and left ductus or left aortic arch with aberrant right subclavian artery and right ductus
    6) pulmonary artery sling
  2. Congenital tumor or cyst of mediastinum
    1) Teratoma
    2) Cystic hygroma, hemangioma
    3) Neurogenic tumor

May I ask some questions about the clinical history at this point? First, did you have any difficulty during endotracheal intubation, and what did you think of the depth and location of the endotracheal tube? Second, if feeding had been started was there any episode of vomiting?

Dr. E. Kim(Sung Ae Hospital); We did not have any difficulty during endotracheal intubation except for the direct laryngoscopic finding of the epiglottis which appeared to be sucked in during inspiration. We also thought that the depth and location of the endotracheal tube were appropriate in this case. This baby was not given formula or breast milk after birth.

Dr. Kim: Thank you, Now may I see the radiologic findings of this newborn infant?

Dr. I. Kim(Radiology): The initial chest X-ray film shows a suspicious right aortic arch, proximal esophageal dilatation, relatively normal heart size and normal pulmonary vascularity and aeration except for focal air trapping in the right middle and lower lung fields. The follow up chest X-ray also shows a suspicious right aortic arch suggestive of aortic arch anomalies. Emphysematous change of the right lung fields was not apparent. After endotracheal intubation, the location of the endotracheal tube was considered appropriate. The feeding tube was located in the lumen of the stomach. Increased heart size by chest X-ray was considered to be an effect of the expiratory phase.

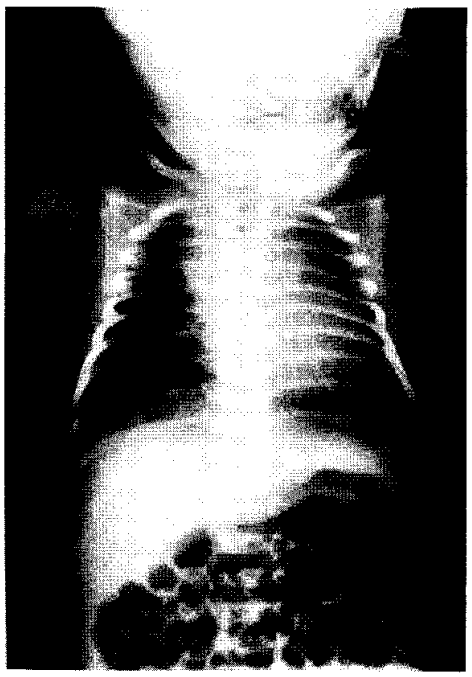

Fig. 1. Chest X-ray at the time of admission to Sung-Ae Hospital, shows right aortic arch and probable right-sided descending aorta. The lungs are aerated and the vascularity is unremarkable.

The nature of continued focalair trapping in the right middle and lower lung fields is not certain. The chest X-ray does not show any mass density.

Dr. B. Kim; Thank you, Dr. Kim. I would like to exclude the possibility of extrinsic tracheal compression due to congenital mediastinal tumor or cyst on the grounds of physical and radiological findings. Development of tracheal cartilage support may rarely be delayed, resulting in tracheomalacia. Many cases of tracheomalacia usually improve by 6 to 12 months of age. Severe cases may benefit from treatment with constant positive airway pressure, and a frequent association with tracheoesophageal fistula has been reported (Hansen and Corbet 1991). So, I would like to exclude the possibility of tracheomalacia in this case. Then there remain two possibilities; congenital tracheal stenosis and vascular ring or sling. In many cases of congenital tracheal stenosis, other assoicated congenital malformations are also present, such as vascular ring anomalies, congenital heart defects, tracheoesophageal fistula, or hemivertebrae (Salzberg and Krumel 1990). In this case, I cannot completely exclude the possibility of congenital tracheal stenosis in spite of no evidence of difficulties during tracheal intubation. Another possibility would be extrinsic obstruction due to vascular ring or sling on the grounds of clinical manifestations and radiologic findings. Here I would like to differentiate the type of vascular ring or sling. In double aortic arch, the respiratory distress begins early, and is in most instances acute and severe because of tight ring formation. The incidence of double aortic arch is more common than the other types of vascular ring or sling. This anomaly has been reported to be more common in females. In right aortic arch with left ligamentum arteriosum or patent ductus arteriosus, or left aortic arch with right ligamentum arteriosum or patent ductus arteriosus, respiratory distress begings later, and is less acute. This anomaly has been reported to be more common in males. In aberrant right subclavian artery, most cases are often asymptomatic because of posterior compression of the esophagus only. In right aortic arch with aberrant left subclavian artery and left ductus, or left aortic arch with aberrant right subclavian artery and right ductus, vascular ring formation is looser than double aortic arch. So respiratory distress is less acute and less severe, and begins later. In anomalous innominate artery or carotid artery, respiratory distress is less acute and less severe, and begins later because of anterior compression of the trachea. In anomalous left pulmonary artery(pulmonary artery sling), incidence is least common among among all types, and asymmetric emphysematous change or atelectasis is accompanied because of compression of the posterior tracheal wall and right or left main bronchus. In the severe form of this anomaly, ring-sling complex and bronchial stenosis are accompanied(Lierl 1993).

In summary, considering the rapidly progressive and fatal respiratory distress and radiologically symmetric aeration, I think that this newborn infant had a double aortic arch with complete vascular ring

Clinical diagnosis(Dr. Kim)

1. Vascular ring. Type: double aortic arch
2. R/O associated tracheal stenosis
3. Laryngomalacia

PATHOLOGIC FINDINGS(Dr. Chi)

At autopsy there indeed was a complete duplication of the aortic arch. Both the right aortic arch and the left aortic arch were patent throughout their entire lengths. There was no focal narrowing of any part of the arch. The diameter of the left arch was greater than that of the right arch. The left ductus arteriosus was larger than the right ductus which was barely patent through the stenotic tract. The diameter of the vascular ring was 0.7cm in maximum extent. It was ovoid and contained trachea and esophagus. The trachea was flattened due to the compression by the vascular ring, barely allowing 0.1 × 0.2cm. The compression level of

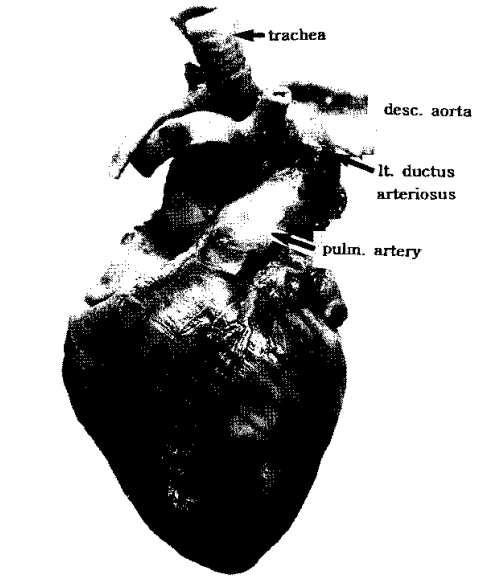

Fig. 2. Frontal view of the removed heart shows a trachea surrounded by double aortic arch associated with a large ductus arteriosus.

Fig. 3. Complete vascular ring shown after the trachea and esophagus are removed. Right and left aortic arches are well shown.

the trachea was approximately 1cm from the tracheal bifurcation. The remaining heart had no intrinsic anomaly. The lungs showed edema and focal alveolar hemorrhage. The liver showed micovesicular fat and extramedullary hemopoiesis. There was early acute tubular necrosis in the kidneys.

Pathologic diagnosis

1. Double aortic arch forming a complete vascular ring with compression of the lower trachea and resultant stenosis
2. Multiple blebs, lungs
3. Recent hemorrhage and erosion, esophagus
4. Petechial hemorrhage, stomach
5. Generalized passive congestion of the viscera
6. Gaseous distention of the gastrointestinal tract
7. Accessory spleen, left paradrenal
8. Focal interstitial hemorrhage, kidney
9. Microvesicular fatty change, liver
10. Extramedullary hemopoiesis, liver
11. Intrathyroidal skeletal muscle bundle
12. Microcytic change, adrenal gland
13. Early acute tubular necrosis, kidney
14. Microcystic change, endocervical gland

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