LEFT PULMONARY ARTERY SLING WITH TRACHEAL STENOSIS

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ABSTRACT

We present the case of a 4-year-old girl, bronchofiberscopy (scope diameter 2.8mm): one third below of trachea, which was divided into two orifices. The orifice on the right side was larger and divided into two suborifices. The left orifice was stenosis and could not pass the bronchoscope. Whole trachea mucosa was red and edema, and could not observe the cartilage ring of trachea. Chest X-ray: infiltration of right lower lobe. Chest CT multiple probes with contrast injection confirmed that: At the sixth to seventh thoracic vertebral level, the left pulmonary artery was arisen from the right pulmonary artery then passed in between the behind trachea and anterior oesophagus to reach the left lung hilar. And at the fourth to fifth thoracic vertebral level, the right upper bronchus was arisen from trachea and then the trachea was shrunk diameter to reach the left lung hilar; at the end, the trachea divided 2 bronchi: left main bronchus and “bridging bronchus”. That bronchus from left lung hilar returned to the right lung.

This patient was diagnosed birth defect: Left pulmonary artery sling with trachea stenosis, type IIA.

Keywords: Congenital malformations of left pulmonary artery, left pulmonary artery sling, tracheal stenosis

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Report: A 4-year-old pediatric patient, admitted on April 24th, 2018 with symptoms of fever, cough, prolonged cloudy sputum.

Pre-history: Child was born full term, often cough, fever, sputum, dysphagia, and choking

Clinical: Patient was alert, had good contact, glasgow 15 points, temperature 36.5°C, weight 15 kg. Pulse 114 times/minute, breathing rate 22 cycles/minute, height 95cm.

After examination, regular heart rate, 114 times/minute, clear heart sound, no pathological sound, symmetrical chest, breathing rate 22 cycles/minute, SpO2 96%, moist rales, dry rales, scatteredly sonorous rales on both sides of the lungs. Other organs did not detect pathological signs.

Subclinical: Blood tests: BC 8.9G/L, N 23.6%, HC 4.4T/L, HST 12.7g/l, TC 431g/L.
Ure 4.2 mmol/l, Cre 45 µmol/l, Na 141 mmol/l, K 4.0 mmol/l, Cl 101 mmol/l, GOT 25 UI/L, GPT 11 UI/L, CRP 0.2mg/l.

Bronchoalveolar lavage: GenXpert (-), TB bacteria culture (-)

Bronchoscopy: one third below of trachea, which was divided into two orifices: right and left. The orifice on the right side was larger and divided into two suborifices. The left orifice was stenosis and could not pass the 2.8 mm bronchoscope. Whole trachea mucosa was red and edema, and could not observe the cartilage ring of trachea.

Diagnosis at admission: bronchopneumonia, patient was treated: antibiotics, anti-inflammatory agent and anti-spasmodic agent. After 15 days of treatment, the patient was discharged from the hospital with alertness, no fever, cough, but still having sonorous rales on both sides of the lungs. Diagnosis determined at discharge:

Normal echocardiography. Straight chest X-ray on May 25th, 2018: Infiltration of right lower lobe (Figure 1). Chest CT multiple probes with contrast injection to construct blood vessels and airways confirmed that: At the sixth to seventh thoracic vertebral level, the left pulmonary artery was arisen from the right pulmonary artery then passed in between the behind trachea and anterior oesophagus to reach the left lung hilar (Figure 2). And at the fourth to fifth thoracic vertebral level (Figure 3), the right upper bronchus was arisen from trachea and then the trachea was shrunk diameter to reach the left lung hilar, at the end, the trachea divided 2 bronchi: left main bronchus and “bridging bronchus”. That bronchus from left lung hilar returned to the right lung (Figure 3).

Diagnosis at admission: bronchopneumonia/airway abnormalities and abnormalities arising from the left pulmonary artery (left pulmonary artery sling with tracheal stenosis type IIA).

Discussion

Left pulmonary artery sling/LPAS is a congenital defect caused by an original external abnormality of the left pulmonary artery; this anomalous artery arises from the right pulmonary artery and then goes from right to left behind the trachea or carina and in front of the esophagus to reach the left lung hilar. On the way, it compresses the lower part of the trachea and bronchus principalis and causes symptoms of upper respiratory tract. This compression can produce left lung emphysema, obstruction, collapse. Therefore, it is important to early recognize and diagnose left pulmonary artery sling defect.

LPAS was first discovered by Glaevecke and Doehle in 1897 [1]. LPAS is very rare, currently even the US and other countries in the world have not had statistics on the incidence of this disease. A retrospective study with statistics from Chinese
hospitals from 2007 to 2014 had 71 cases of LPAS out of 52,200 patients with congenital heart disease, accounting for 0.014% [2]. LPAS is often associated with tracheobronchial, heart defects and other non-cardiac abnormalities [3,4]. Defects of airways in association with LPAS include diseases: incomplete tracheobronchial cartilage, tracheobronchial cartilage cordis, tracheal segment stenosis, and these associated defects increase airway obstruction.

According to Well et al. [3], a combination of 32 studies of LPAS found heart defect associated with LPAS was about 50% of cases, most defects were associated with atrial septal defect, patent ductus arteriosus, ventricular septal defect and left superior vena cava. Other defects in association with LPAS include: imperforate anus, biliary tract atrophy, genital defect, ovary defect and lung parenchyma thyroid gland.

According to Well et al., patients with LPAS have 50% of associated congenital tracheal stenosis. Based on their study results, the author suggested LPAS be divided into 2 main types: type 1 and type 2, the division of these 2 types was based on the position of the carina in the chest: type I carina at the fourth to fifth thoracic vertebral level, type II carina at the lower position, sixth to seventh thoracic vertebral level. These two types were further subdivided into two subtypes based on: upper lobe bronchus or no right upper lobe bronchus: type II A with upper lobe bronchus and type II B without right upper lobe bronchus (Figure 4).

Figure 4. Diagram of classification of left pulmonary artery defects

Our patient presented left pulmonary artery sling with tracheal stenosis/LPAS Type IIA and no other defects were detected.

Pathogenesis: When the left pulmonary artery passes behind, the end of the right and posterior left bronchus principalis of the trachea compresses the right trachea and bronchus, causing a major effect on the right lung and also obstructing the two sides. This anomalous artery supplies blood to one side or both sides of the lungs, often anomalous artery only supplies blood to the left upper lobe, and the right pulmonary artery supplies blood to the left lower lobe; in addition, a partial anomalous artery supplies blood to the right upper lobe [5].

Complications: Because LPAS is associated with tracheobronchial stenosis, the symptoms of airway obstruction often occurred. Patient reported had a recurrence of respiratory infections many times a year.

Prognosis: Early diagnosis led to good prognosis, surgical mortality increased in patients with associated airway stenosis. Long-term surgical and follow-up patients showed good prognosis [5].

Pre-history and clinical signs: patients with LPAS for the first few weeks of life often showed signs of respiratory distress, stridor, cyanosis, wheezing or pneumonia. Clinical examination found respiratory distress, stridor, wheezing. If there was a tracheal obstruction, patients showed symptoms of fast shortness of breath, chest shrinkage when treated but fixed and if there were more associated congenital heart defects, then typical clinical symptoms above can be expressed more fully and clearly [5].

Patient we reported had LPAS with tracheal stenosis, so signs of wheezing appeared continuously...
and fixed in the two lungs, disappeared after 15 days of treatment with antibiotics, anti-inflammatory agent and antispasmodic agent.

Diagnosis set up to determine the LPAS with tracheal stenosis must be based on a number of image diagnostic and bronchoscopy techniques [5]:

- Image diagnosis
  + Chest X-ray: the trachea was lower than normal, deflected to the left and may be compressed to the right, increased the right lung ventilation due to compression of the right bronchus principalis. The left lung can also show increased ventilation due to compression in the position where divides the carina and the left bronchus principalis. Patients with severe obstruction showed a collapse of one side of the lung or lobe of the lung.

  Patient we reported had lower trachea, dividing the carina at the sixth to seventh thoracic vertebral level, tracheal stenosis caused infiltration of right lower lobe.

  + Barium-containing esophagography is often a recommended diagnostic technique; injury was a frontal esophagus indentation due to compression of LPAS.

  + Echocardiography: no pulmonary artery trunk divided the right pulmonary artery and the left pulmonary artery, no left pulmonary artery arose from the right pulmonary artery then passed in between the behind trachea and anterior oesophagus to reach the left lung hilar. In addition, echocardiography also detected other heart defects.

  + MRI: Chest magnetic resonance imaging, vascular magnetic resonance imaging, chest CT scan or a combination of three these techniques can describe details of anatomy as well as three-dimensional reconstruction of LPAS anatomy and airway injury (1)

- Bronchoscopy: For LPAS alone, bronchoscopy is not necessary, but if there are signs of tracheal compression, tracheal stenosis, bronchoscopy is necessary to assess the injury of associated airway defect [6]. Differential diagnosis: left pulmonary artery sling with tracheal stenosis should be differential diagnosed with other diseases with clinical symptoms similar to this disease: aortic double arch, mediastinal tumor, tracheal stenosis, tracheal cartilage cords and stridor.

**Treatment**

LPAS alone needs to be treated to regenerate the left pulmonary artery by cutting off the abnormal artery connecting to the pulmonary artery trunk. In case with tracheal stenosis, shaping trachea is needed. Associated tracheal shaping surgery is very complicated and has severe prognosis, so only tracheal shaping is performed in patients with clear clinical symptoms due to airway obstruction (7).

**Conclusion**

Left pulmonary artery sling with tracheal stenosis is a rare congenital disease, the diagnosis of this disease is mainly based on chest CT multiple probes, echocardiography, chest MRI, vascular MRI for details of anatomy of this abnormal pulmonary artery. The disease should be early detected for timely treatment, surgery for reconstruction of left pulmonary artery and airway is the best treatment. Patient we reported needs to have surgery for reconstruction of left pulmonary artery and airway.

**REFERENCES**


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