J Korean Med Sci 2000; 15: 119-21
ISSN 1011-8934

Unusual Association of Pulmonary Artery Sling with Right Aortic Arch and Aberrant Left Subclavian Artery

We present an unusual case of vascular sling, tracheal stenosis by complete cartilaginous ring, and aberrant left subclavian artery with right aortic arch that underwent successful surgical repair for the sling. These abnormalities were suspected from unusual multiple indentations found on esophagogram. Complete preoperative diagnosis was established with chest computerized tomogram combined with angiography.

Key Words: *Tracheal Stenosis; Subclavian Artery; Aorta*

Dong Ho Jeong, Heon-Seok Han, Youn-Soo Hahn, Seog-Jae Lee*

Departments of Pediatrics and Thoracic Surgery*, Chungbuk National University College of Medicine, Cheongju, Korea

Received: 26 May 1999
Accepted: 12 July 1999

Address for correspondence
Heon-Seok Han, M.D.
Department of Pediatrics, Chungbuk National University College of Medicine, 42 Gaesin-dong, Heungduk-gu, Cheongju 361-711, Korea
Tel: +82.431-269-6370, Fax: +82.431-264-6620
E-mail: hshan@med.chungbuk.ac.kr

INTRODUCTION

The origin of the left pulmonary artery from the right pulmonary artery, also known as anomalous left pulmonary artery, or pulmonary artery sling, is a rare anomaly in which the lower trachea is partially surrounded by vascular structure (1). This anomaly is frequently associated with complete cartilaginous ring in the distal trachea, resulting in tracheal stenosis. The association of pulmonary artery sling with aortic arch anomaly such as right aortic arch or aberrant subclavian artery is very rare. When the right aortic arch with retroesophageal aberrant left subclavian artery has no left-sided ductus arteriosus or ligamentum, vascular ring does not form and no treatment is required. We describe here an unusual combination of vascular sling, tracheal stenosis by complete cartilaginous ring, and aberrant left subclavian artery with right aortic arch.

CASE REPORT

A 7-year-old boy presented with complaints of wheezing and intermittent dyspnea since six months of age. He was treated as having bronchiolitis and bronchial asthma with intermittent medications. Recently he developed slightly decreased exercise tolerance. Chest roentgenogram showed hyperinflated lung field and normal cardiac silhouette. Electrocardiogram was normal. No abnormal finding was noted on allergy test. His trachea was diffusely narrowed on endoscopy. Three-dimensional computerized tomogram (CT) showed low carina, horizontal mainstem bronchi, and long-segment tracheal stenosis (Fig. 1A). Multiple indented areas were observed in the esophagogram (Fig. 1B-C). Each indentation was confirmed by the subsequent cardiovascular studies. Angiography confirmed right aortic arch, right-sided descending aorta, and aberrant left subclavian artery arising from left side of descending aorta (Fig. 2A). Chest computerized tomogram revealed complete cartilaginous ring as a cause of diffuse tracheal narrowing, retroesophageal course of left subclavian artery (Fig. 2B), and left pulmonary artery arising from right pulmonary artery forming vascular sling compressing trachea (Fig. 2C). Retrospectively, multiple esophageal indentations were identified as right aortic arch, aberrant left subclavian artery, and vascular sling. Surgical correction for the vascular sling was carried out using standard cardio-pulmonary bypass techniques. Left pulmonary artery was dissected from its site of origin and implanted to the main pulmonary artery anterior to trachea. Distal trachea was maintained as round form without narrow point. He made a smooth post-operative recovery and is in good health with only a mild wheezing sound.

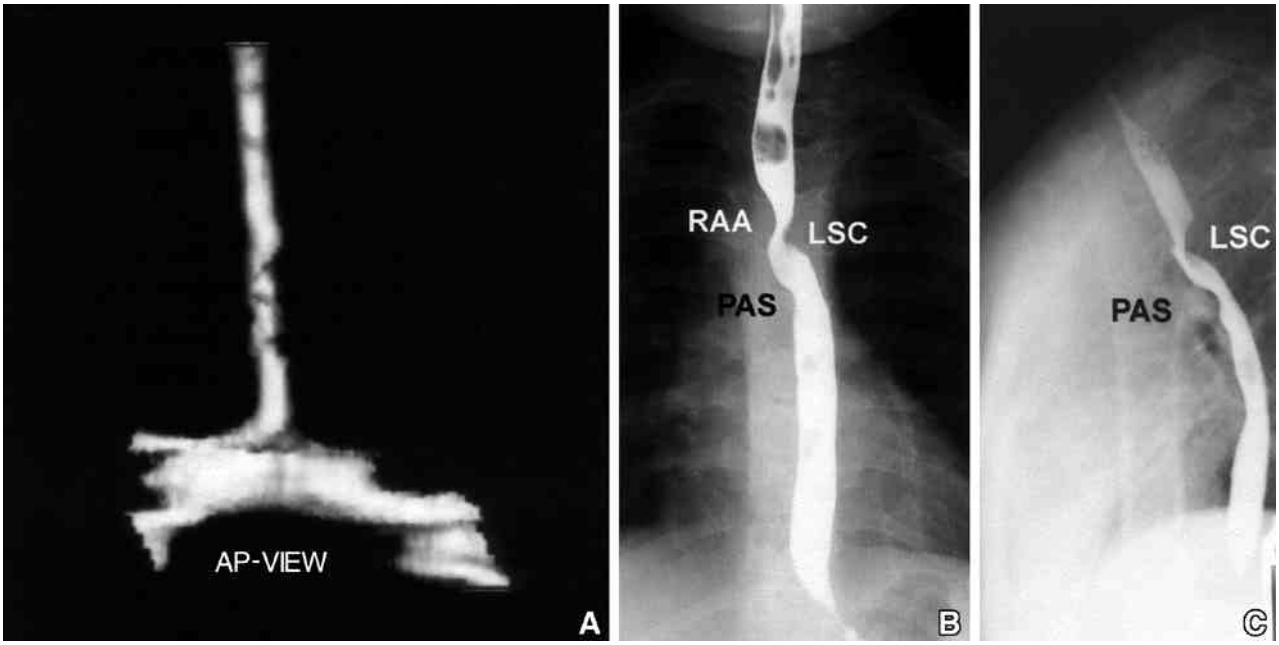

Fig. 1. A: Three-dimensional computerized tomogram of trachea shows long-segment tracheal stenosis, low carina, and horizontal mainstem bronchi. **B:** Multiple narrow points of esophagus in AP esophagogram: right aortic arch (RAA); pulmonary artery sling (PAS); aberrant left subclavian artery (LSC). **C:** Lateral esophagogram shows multiple narrow points: pulmonary artery sling (PAS); aberrant left subclavian artery (LSC).

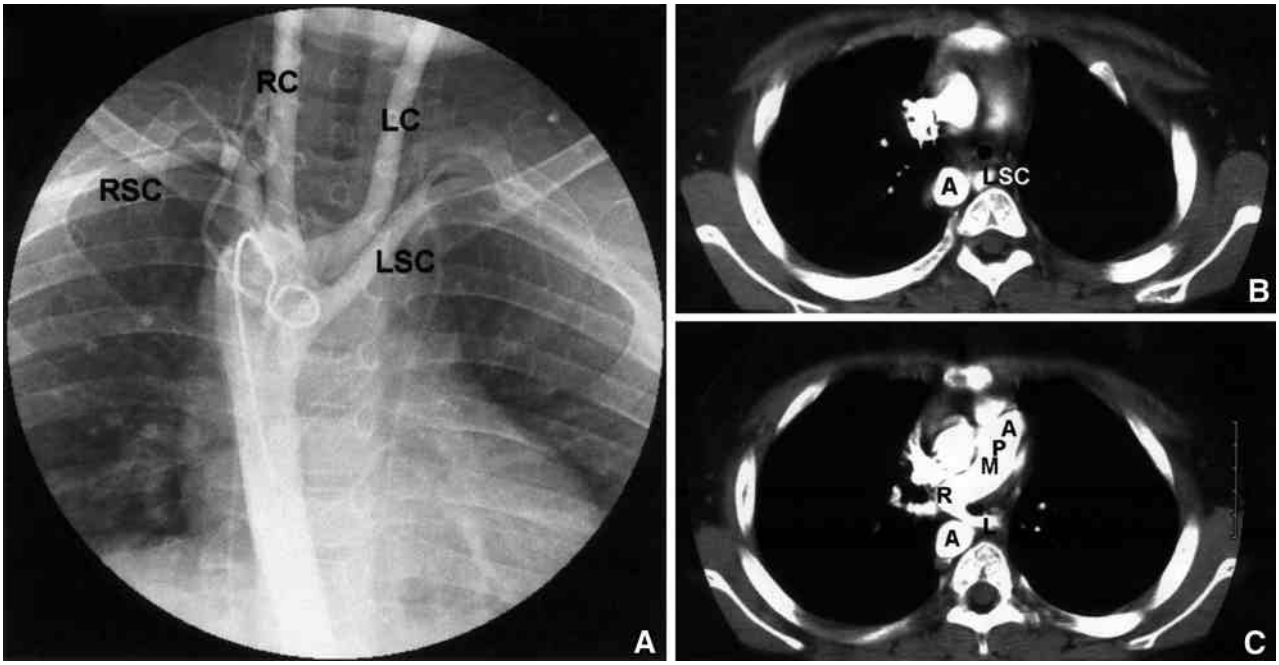

Fig. 2. A: Aortogram shows right aortic arch, right descending aorta, and aberrant left subclavian artery course. The sequence of brachiocephalic arteries are left carotid (LC), right carotid (RC), right subclavian (RSC), and retroesophageal left subclavian (LSC). **B:** Computerized tomogram of chest shows complete cartilaginous ring as a cause of tracheal narrowing in front of aberrant left subclavian artery (LSC) arising from the left side of descending aorta (A). **C:** Chest CT shows that left pulmonary artery (L) arises from right pulmonary artery (R), forming a vascular sling that compresses the trachea.

DISCUSSION

Those patients with pulmonary artery sling typically present with severe respiratory distress and stridor, although milder forms do exist and may be identified incidentally during angiography for another cardiovascular

anomaly (1). Persistent high mortality in infants with pulmonary artery sling (retrotracheal anomalous left pulmonary artery) is primarily due to the co-existence of long-segment tracheal stenosis due to complete cartilage rings. Berdon used the term ring-sling complex for this frequent association (2). In our case, the patient's main complaint was caused by tracheal stenosis and vascular sling, rather than aberrant left subclavian artery. There was no ductus or ligamentum forming vascular ring with aberrant left subclavian artery. No procedures was performed for the aberrant subclavian artery. The tracheal stenosis was thought to be mild, because he survived infancy and presented persistent asthmatic symptom.

Beekman suggested that magnetic resonance imaging (MRI) should be performed for patients suspected of having a vascular cause for stridor or dysphagia. If there is a need for a screening procedure, color Doppler echocardiography should be used and if the results of that are equivocal or non-conclusive, esophagography and bronchoscopy should be used. If MRI is difficult to interpret, it should be augmented by magnetic resonance angiography (MRA) before considering cine-angiography (3). A report from the Mayo Clinic also suggested magnetic resonance imaging as the technique of choice for the delineation of the vascular and tracheal anatomy in patients suspected of having a vascular ring after analyzing 39 cases of vascular ring during 46 years (4). MR imaging was useful in the diagnosis and management of patients with airway obstruction related to vascular compression without the need for more invasive procedure (5).

Vascular ring by aberrant subclavian artery can be suspected when normal branching of innominate artery is not seen on echocardiography (right-sided innominate artery in the case of left arch and left-sided innominate artery in the case of right arch). Precordial short axis and subcostal views can also suspect pulmonary artery slings. Even with the systematic echocardiographic approach, several limitations exist, such as defining anterior compression of the trachea, and resolution of every segment of a complete vascular ring (6). We tried to view an aberrant subclavian artery and sling with echocardiography, but the window for suprasternal and subcostal views were not clear enough to delineate the structures.

Among the vascular anomalies causing tracheobronchial compression, patients with innominate artery compression and pulmonary artery sling recover well soon after surgery with relief of most of their symptoms (7).

Our patient's respiratory symptoms were thought to be caused by intrinsic airway narrowing and pulmonary artery sling. We determined to manage only the vascular sling, because the trachea above and below the vascular sling site was wider than the compressed area (figures not shown) in spite of diffuse tracheal stenosis.

The association of pulmonary artery sling with aortic arch anomaly is very rare. Wittenborg described a case of pulmonary artery sling with aberrant right subclavian artery from left aortic arch (8). Jue described three associated aortic arch anomalies in 23 cases of pulmonary sling, which were coarctation, anomalous right subclavian artery and right aortic arch (9). In review of Dohlemann et al. (10), only one out of 115 cases had aberrant right subclavian artery, which was Wittenborg case. Our case is similar to Wittenborg case, but the difference is position of aortic arch and aberrant subclavian artery.

REFERENCES

1. Weinberg PM. *Aortic arch anomalies. In: Emmanouilides GC, Allen HD, Rienmentschneider TA, Gutgesell HP, eds. Heart disease in infants, children, and adolescents. Baltimore: Williams & Wilkins, 1995; 810-37.*
2. Berdon WE, Baker DH, Wung J, Chrispin A, Kozlowski K, de Silva M, Bales P, Alford B. *Complete cartilage-ring tracheal stenosis associated with anomalous left pulmonary artery: the ring-sling complex. Radiology 1984; 152: 57-64.*
3. Beekman RP, Hazekamp MG, Meijboom EJ, de Roos A, Staalman CR, Beek FJA, Ottenkamp J. *A new diagnostic approach to vascular rings and pulmonary slings: the role of MRI. Magnetic Resonance Imaging 1998; 16: 137-45.*
4. van Son JAM, Julsrud PR, Hagler DJ, Sim EKW, Puga FJ, Schaff HV, Danielson GK. *Imaging strategies for vascular rings. Ann Thorac Surg 1994; 57: 604-10.*
5. Mahboubi S, Meyer JS, Hubbard AM, Harty MP, Weinberg PM. *Magnetic resonance imaging of airway obstruction resulting from vascular anomalies. Int J Pediatr Otorhinolaryngol 1994; 28: 111-23.*
6. Parikh SR, Ensing GJ, Darragh RK, Caldwell RL. *Rings, slings and such things: diagnosis and management with special emphasis on the role of echocardiography. J Am Soc Echocardiogr 1993; 6: 1-11.*
7. Anand R, Dooley KJ, Williams WH, Vincent RN. *Follow-up of surgical correction of vascular anomalies causing tracheobronchial compression. Pediatr Cardiol 1994; 15: 58-61.*
8. Wittenborg MH, Tantiwongse T, Rosenberg BF. *Anomalous course of left pulmonary artery with respiratory obstruction. Radiology 1956; 67: 339-45.*
9. Jue K, Raghib G, Amplatz K, Adams P, Edwards JE. *Anomalous origin of the left pulmonary artery from the right pulmonary artery. Am J Roentgenol 1965; 95: 598-610.*
10. Dohlemann C, Mantel K, Vogl TJ, Nicolai T, Schneider K, Hammerer I, Apitz J, Meisner H, Joppich I. *Pulmonary sling: morphological findings. Pre- and postoperative course. Eur J Pediatr 1995; 154: 2-14.*