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A Rare Case of Tracheomalacia Associated with Vascular Ring in an Infant

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Tracheomalacia can be a life threatening upper air way obstructive disease in an infant and vascular rings can be also a major rare cause of tracheoesophageal obstruction. These two rare entities can be combined in one patient because the vascular ring can cause secondary tracheomalacia during development of fetus. The diagnosis of this combination and adequate surgical correction is occasionally difficult. This is a report of an infant who had not diagnosed tracheomalacia associated with vascular ring until 5 months of age because of the prolonged tracheal intubation. The rigid bronchoscopic examination performed under impression of tracheomalacia revealed a concentric tracheal collapse, an unusual bronchoscopic findings of tracheomalacia, which raised a suspicion of the tracheal compression by vascular rings. The 3-D reconstructive CT aortography clearly demonstrated the double aortic arch. The patient was treated surgically by simple division of the left aortic arch and aortopexy with good result. The vascular ring such as double aortic arch should be considered during the diagnosis of tracheomalacia in infants. If the tracheomalacia is associated with vascular ring, simultaneous surgical correction should be performed.

Key Words : Tracheomalacia, Vascular ring, Double aortic arch, Infant

I. INTRODUCTION

Tracheomalacia is a disorder that occurs almost exclusively in infancy and early childhood that results in a soft "floppy" or "flaccid" trachea form abnormal cartilage development. Severe tracheomalacia has been implicated in case of sudden infant death syndrome. This dying anoxic spell can be relived by aortopexy in modern pediatric surgery. Although tracheomalacia is most frequently associated with esophageal atresia with tracheoesophageal fistula, it is rarely associated with vascular ring. The vascular ring can cause direct tracheoesophageal compression and the secondary dysplastic change of the developing tracheal cartilage. In tracheomalacia associated with vascular ring, aortopexy or division of vascular ring alone may not always relive the air way obstruction completely. In this situation, concomitant aortopexy is safe policy to treat the all underlying problems. This case report emphasizes the importance of the preoperative recognition and one stage surgical correction in the management of tracheomalacia and vascular ring.

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II. CASE REPORT

A 3.5 Kg male baby was born by spontaneous vaginal delivery at 40 weeks gestation. He had done well without problem until the 20thday of life when he developed sudden cyanotic anoxic spell during bottle-feeding. He was intubated, ventilated, and admitted in another hospital. During the 3 months of admission, he had failed several attempts of extubation because of immediate respiratory distress after extubation. The fluoroscopic X-ray examination of the tracheobrochial treeshowed the collapse of the intrathoracic trachea during exhalation. The tentative diagnosis of tracheomalacia was made and transferred to our hospital. At the time of transfer, the baby was a healthy 4-months-old infant except the intubated status. Adequate ventilation was possible only when the tip of the tracheal tube was located at between the collapsed part of trachea and the carina. The echocardiography showed patent ductus arterosus (PDA). The 3-D reconstructive CT tracheography showed the segmental collapse of the lower third of trachea (Fig. 1-A). The rigid bronchoscopic examination with spontaneous ventilation confirmed the tracheomalacia. However, an atypical bronchoscopic findings, the pulsatile concentric collapse of trachea, was observed instead of the typical bronchoscopic findings of tracheomalacia (Fig. 2). The esophagography also depicted the narrowing of the midesophagus (Fig. 3-A). The both the concentric pulsatile collapse of trachea and the narrowing of the esophagus led us suspect

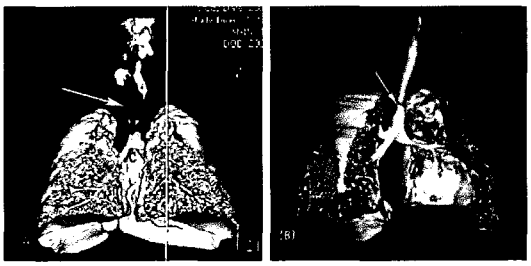

Fig. 1. (A) The preoperative 3-D reconstructive CT tracheography showed the segmental collapse of the lower third of trachea (arrow). (B) The postoperative 3-D reconstructive CT tracheography showed disappearance of the collapsed segmental of trachea with minimal residuum (arrow).
Abbreviations: C; carina

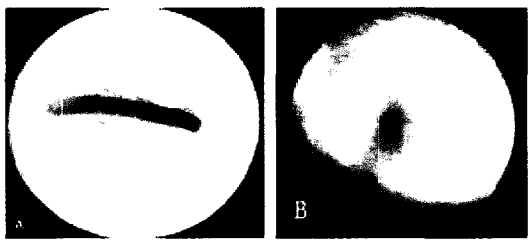

Fig. 2. (A) The typical bronchoscopic finding of tracheomalacia is the elliptical collapse of malatic segment of trachea during exhalation. (B) In this case, an atypical pulsatile concentric collapse of trachea was observed in the bronchoscopic examination.

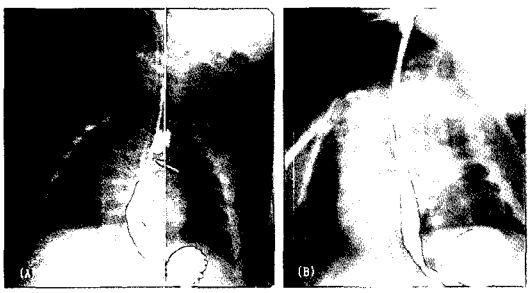

Fig. 3. (A) The preoperative esophagographyshowed the narrowing of the midesophagus (arrow). (B) The postoperative follow-up esophagography revealed the disappearance of the narrowing segment of esophagus.

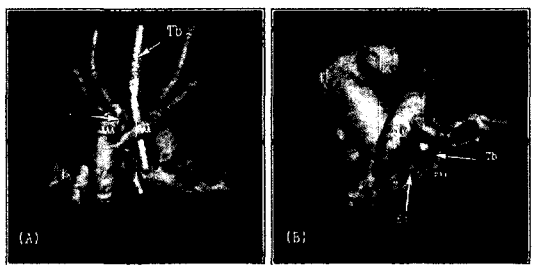

Fig. 4. (A) The 3-D reconstructive CT aortography revealed that two aortic arches encircle the trachea and esophagus. (B) The dominant aortic arch was the anterior aortic arch.
Abbreviations: Es; esophagus, Tb; Intubated trachea, AAA; anterior aortic arch, PAA; posterior aortic arch

the patient to have a tracheomalacia associated with tracheoesophageal compression by a vascular ring. Under the suspect of the vascular ring, the 3-D reconstructive CT aortography was taken, and it showed that two aortic arches encircle the trachea and esophagus (Fig. 4-A).The dominant aortic arch was the anterior aortic arch (Fig. 4-B).

The operation was performed at 127 days of age. A left posterolateral thoracotomy was performed. The left lobe of thymus was removed in order to expose the aortic arch. The PDA was then ligated and divided, as was the posterior aortic arch between the left common carotid and left subclavian arteries. This left the esophagus and trachea free from the vascular compression. The aortopexy was performed under guide of intraoperative bronchoscopic examination. The patient had a good hemodynamic state during surgery. Postoperative recovery was rapid, and the extubation was achieved 4 days later. The patient was significantly improved and discharged from the hospital 11 days later. The clinical and radiographic follow-up examinations demonstrate good results (Fig. 1-B, Fig. 3-B). Follow-up to 18months shows good growth with no respiratory symptoms.

III. DISCUSSION

The tracheomalacia is an uncommon condition in which the tracheal wall is especially soft and pliable. The condition may occur as an isolated anomaly but is most often associated with esophageal atresia and tracheoesophageal fistula. It also can occur in association with other lesions, such as vascular ring and tumor that compress the developing airway. The reported incidence of the vascular ring ranges between 0.3-0.6 percent. The clinical features of vascular ring are asymptomatic, dysphagia and dyspnea. The dysphagia is caused by the partial obstruction of esophagus by the vascular ring, and the respiratory difficulty can be caused by the static or dynamic compression of the airways by vascular ring. Such congenital vascular compression also can cause the secondary change of developing tracheal cartilage, such as hypoplasia, dysplasia, or absence of the normal cartilage framework. All these developmental changes of tracheal frame can result in the tracheomalacia. The other proposed mechanisms for respiratory tract symptoms in vascular ring include intravascular volume infusion, aspiration, exercise-induced dilatation of the aortic arch and age-dependent changes in thoracic compliance.[2)]

The double aortic arch is the most common type of the vascular ring anomaly and the major cause of tracheoesophageal obstruction in vascular ring.[2)] The normal formation of the aortic arch requires resorption of the dorsal fourth right arch with remodeling of the right subclavian and common carotid into an innominate artery arising as the first of the arch. The double aortic arch results when this resorptive process does not occur properly. Although the tracheomalacia in association with the vascular ringis a very rare condition, it should be always considered the concurrence of two entities in one patient.

It is very important the meticulous diagnostic workup to delineate the anatomic factors causing the air way obstruction, as well as to determine the adequate surgical strategies to relieve it. The mainstays of initial evaluation under suspicion of the airway anomaly in infants and children are chest radiography, cine air-tracheography, esophagography and rigid bronchoscope. The echocardiography also should be done for exclusion of other congenital cardiac anomalies and as the screening study of the associated vascular ring in diagnosis of tracheomalacia. The esoph-

agography is also useful study to reveal the associated congenital esophageal anomalies and the vascular compression of esophagus. The bronchoscopic examination is the main diagnostic study to confirm the tracheomalacia. We emphasize that the bronchoscopic findings in our case. It showed the peculiar finings such as concentric pulsating collapse of tracheal wall. These findings raised us a suspicion of the tracheal compression by vascular rings. Chest CT imaging with three dimensional reconstructions enable full evaluation of the vascular anatomy in our case. Three dimensional imaging may aid in diagnosis of innominate artery compression syndrome by demonstrating the extent of the tracheal luminal narrowing, the tracheal configuration, the structure causing the compression, and the size of the thymus. It is very useful in assessing relationships between vascular structures and the adjacent trachea, and alsouseful in assessing and monitoring the surgical result.

Most children with tracheomalacia can be treated conservatively because the natural history of the condition is significant spontaneous improvement after age of one year.[3] Surgicalcorrection should be reserved for patients with life threatening attacks, those who cannot be extubated, and those who have been repeated bouts of pneumonia.[4] Aortopexy is recognized as the standard surgery for tracheomalacia.[5] The aim of aortopexy is to provide sufficient space in the mediastinum so that the malatic trachea will not be compressed by the adjacent vascular structures or the esophagus. However, the optimal managementof tracheomalacia associated with vascular ring is ill defined, particularly in the very young infants. A degree of tracheomalacia may be responsible for some residual symptoms after division of vascular ring alone.[6] Two case reports are presented in children who developed significant respiratory distress due to persistent tracheomalacia despite vascular decompression for double aortic arch.[7] Some children died from complications of a severe tracheomalacia after vascular division alone.[8)~10)] Aortopexy was commonly used to further alleviate compression and the in tendency for tracheal collapseafter division of vascular ring.[11)12)] Many reports support that the management requires division of the vascular ring and, if tracheomalacia is present, some form of aortopexy.[6)~13)]

We emphasize the importance of the preoperative detection of two entities, "the vascular ring anomaly and the tracheomalacia" and the importance of the concomitant correction of both of them.

References

1. Cordovilla ZG, Cabo SJ, Sanz GE, Moreno GF, Alvarez DF. *Vascular rings of aortic origin: the surgical experience in 43 cases. Rev Esp Cardiol 1994;7:468-75.*
2. Grathwohl KW, Afifi AY, Dillard TA, Olson JP, Heric BR. *Vascular rings of the thoracic aorta in adults. Am Surg 1999;65:1077-83.*
3. Kiley EM, Spitz L, Brereton R: *Management of tracheomalacia by aortopexy. Pediatr Surg Int 1987;2:13-16.*
4. Filler RE, Messino A, Vinograd I: *Severe tracheomalacia associated with esophageal atresia: results of surgical treatment. J Pediatr Surg 1992; 27:1136-1141.*
5. Benjamin B: *Tracheomalacia in infants and children . Ann Otol Rhino Laryngol 1984;93:438-446.*
6. Han MT, Hall DG, Manche A, Rittenhouse EA. *Double aortic arch causing tracheoesophageal compression. Am J Surg 1993;165:628-31.*
7. Colquhoun IW, Pollock JC. *Distal tracheal stenosis in two children with double aortic arch and associated tracheomalacia. Eur J Cardiothorac*

Surg 1990;4:287-9.

8. Maksoud Filho JG, Goncalves ME, Tannuri U, Maksoud JG. *Compression of esophagus and trachea by aortic arch anomalies in childhood. Rev Assoc Med Bras 199339:165-9.*
9. Horvath P, Hucin B, Hruda J, Sulc J, Brezovsky P, Tuma S, et al: *Intermediate to late results of surgical relief of vascular tracheobronchial compression. Eur J Cardiothorac Surg 1992;6: 366-71.*
10. Rivilla F, Utrilla JG, Alvarez F. *Surgical Management and follow-up of vascular rings. Z Kinderchir 1989;44:199-202.*
11. Becker CL, Ilbawi MN, Idriss FS, DeLeon SY. *Vascular anomalies causing tracheoesophagealcompression. J Thorac Cardiovasc Surg 1989;97:725-731.*
12. Robert CS, Otterson HB, Sade RM, Smith CP III, Tagge EP, et al. *Tracheoesophageal compression from aortic arch anomalies. Analysis of 30 operatively treated children. J Ped Surg 1994;29:334-338.*
13. Sebening C, Jakob H, Tochtermann U, Lange R, Vahl CF, Bodegom P, et al. *Vascular tracheobronchial compression syndromes-experience in surgical treatment and literature review. Thorac Cardiovasc Surg 2000 48164-74.*