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Complete Vascular Ring Caused by Kommerell's Diverticulum and Right Aortic Arch with Mirror Image Branching

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Vascular ring, caused by Kommerell's diverticulum and ligamentum arteriosum, in a patient with right aortic arch and mirror image branching is extremely rare. A 10-month-old boy with coughing and stridor was diagnosed as having tracheo-esophageal stenosis, which is caused by a vascular ring with Kommerell's diverticulum, ligamentum arteriosum, right aortic arch, and mirror image branching. Kommerell's diverticulum was successfully resected via a left thoracotomy. The patient has been free from tracheo-esophageal stenosis for a year after the surgery.

Key words: 1. Aorta arch
2. Vascular ring
3. Diverticulum

CASE REPORT

A 10-month-old male child was presented at Dankook University Hospital with coughing and grunting. This child was born on 32nd week of the gestational period. He weighed 1,838 g at birth, and underwent neonatal intensive care for a month. The child had been diagnosed as having a vascular ring, during hospitalization for the treatment of bronchiolitis, seven months after birth. Thereafter he has been managed at the pediatric outpatient clinic. However, despite medical treatment, his respiratory symptoms did not significantly improve. He was admitted again to have the vascular ring treated surgically. The patient weighed 8.7 kg and showed good activity at the time of admission. He exhibited postprandial vomiting a few times per day. The sound of his breathing was coarse and stridor in auscultation.

Preoperative 2-dimensional (2D) echocardiography revealed a right aortic arch with left innominate artery, and Kommerell's diverticulum at the aortic isthmus. There was no intracardiac anomaly. Preoperative esophagography showed a posterior indentation at the upper thoracic esophagus, and this indentation caused severe esophageal stricture (Fig. 1). Preoperative computed tomography (CT) scan revealed right aortic arch, mirror image branching of the arch vessels, Kommerell's diverticulum at isthmic portion of the descending aorta, and focal narrowings of the trachea and esophagus, just above the carina, due to Kommerell's diverticulum (Fig. 2). Preoperative magnetic resonance image showed the ligamentum arteriosum, between Kommerell's diverticulum and the left pulmonary artery. This finding suggested a complete vascular ring.

The surgery was planned to resect Kommerell's diverticulum and liamentum arteriosum. A left lateral thoracotomy was performed, through the fourth intercostal space, under general anesthesia. There was a conical shaped Kommerell's diverticulum at the left side wall of the descending thoracic

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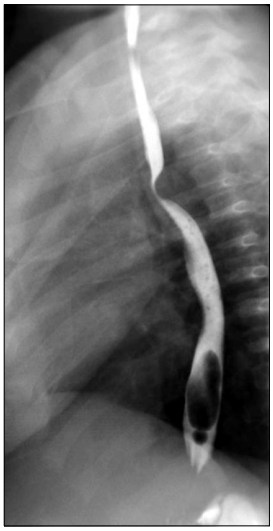

Fig. 1. Preoperative esophagography shows posterior indentation of upper thoracic esophagus.

aorta. The size of Kommerell's diverticulum was 12 mm in diameter, and 10 mm in length. Dense fibrous band appeared to be a ligamentum arteriosum, which had been connected between the conical end of Kommerell's diverticulum and the left pulmonary artery. The ligamentum arteriosum completed the vascular ring. Esophagus was positioned inside the vascular ring, and was severely tightened by the ligamentum arteriosum and Kommerell's diverticulum.

The ligamentum aretiosum was isolated and divided. During the process, the thoracic duct had ruptured, and it was ligated by using surgical clips. Upon managing the rupture, Kommerell's diverticulum and thoracic aorta were dissected, and then Kommerell's diverticulum was resected after a side clamping of the thoracic aorta, and the aortic stump was repaired by using a 6-0 polypropylene suture. During the dissections of ligamentum arteriosum, Kommerell's diverticulum, and the thoracic aorta, the electrocautery was avoided to prevent potential injuries of recurrent laryngeal nerve.

There were no significant postoperative events. Coughing

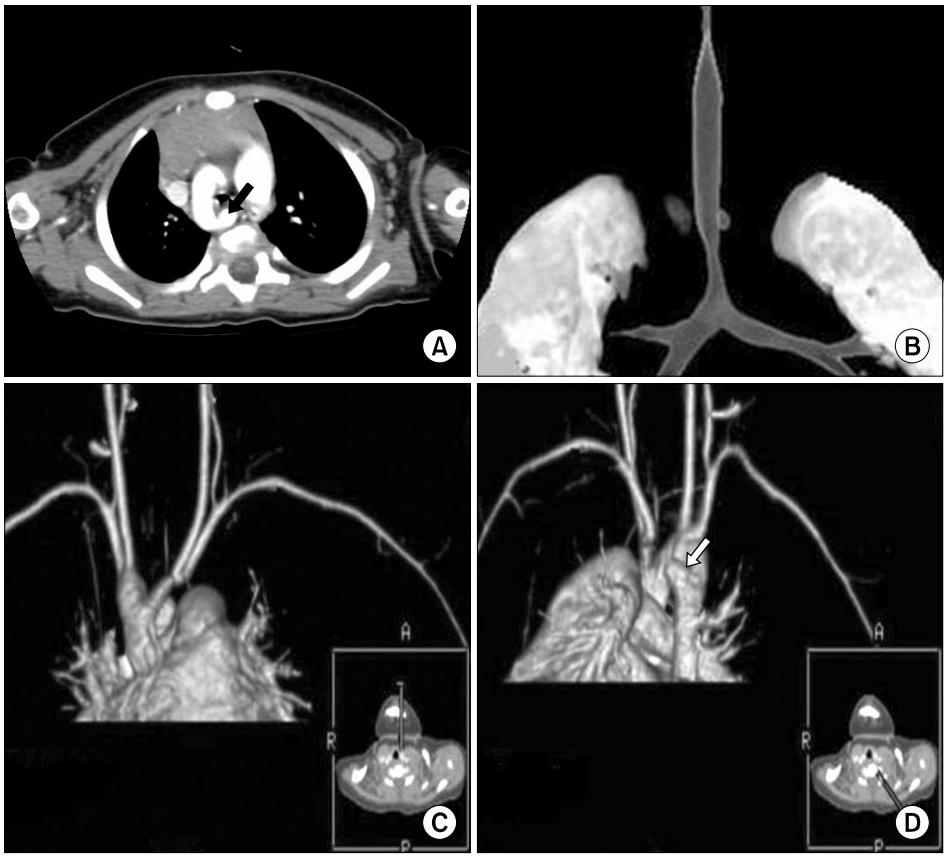

Fig. 2. Preoperative computed tomography scans. (A) Kommerell's diverticulum (black arrow) originated from isthmic portion of the right aortic arch. (B) 3-Dimensional image of trachea shows tracheal stenosis above carina. (C) Anterior view of the aortic arch and branches shows a mirror-image branching pattern. (D) Posterior view of aortic arch shows Kommerell' diverticulum (white arrow) at aortic isthmus.

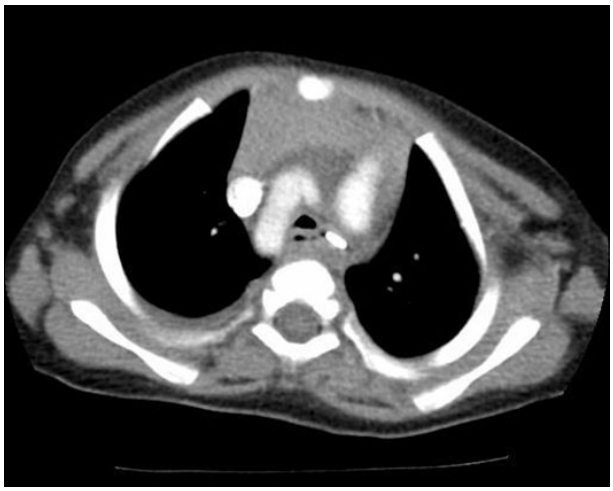

Fig. 3. Postoperative computed tomography scan shows markedly alleviated Kommerell's diverticulum and surgical clip.

and grunting subsided and stridor was no longer heard. Postoperative CT scan showed markedly alleviated Kommerell's diverticulum (Fig. 3). The patient had been discharged 11 days after the operation, and displayed a healthy condition without any symptoms associated with tracheo-esophageal compression for a one-year follow-up.

DISCUSSION

The complete vascular ring is caused by abnormalities of the aortic arch and great arteries, and these abnormalities can give rise to compressed trachea and esophagus [1]. The two most common types of complete vascular rings involve the double aortic arch and right aortic arch with left ligamentum arteriosum [2]. In the right aortic arch with left ligamentum arteriosum, the left subclavian arteries usually originate from an aberrant position. However, a small number of left subclavian arteries originate from a normal position, also called a mirror-image arch branching, as in this case.

During the development of the aorta, the right dorsal aorta remains patent and the left dorsal aorta including left 4th aortic arch, is interrupted abnormally, which could produce a right aortic arch. If the left distal aortic arch, distal to origin of the left subclavian artery, disappears, a mirror image banching is developed as a result [3]. Complete vascular rings, caused by the right aortic arch with mirror image branching, are extremely rare [4].

Kommerell's diverticulm was first described by Burckhard F. Kommerell in 1936 [5]. The definition of Kommerell's diverticulum is a saccular aneurysmal dilatation, which had been found at the origin of an aberrant right subclavian artery or aberrant left subclavian artery [6]. Embryologically, it originates from the persistent distal end of the interrupted 4th aortic arch [5].

In cases of a vascular ring with Kommerell's diverticulum, aberrant subclavian artery or ligamentum arteriosum have a significant role in completing a vascular ring. Kommerell's diverticulum, by itself, has a space-occupying effect. As such, it can be an additional aggravating factor for the compression of the trachea and esophagus. Therefore, Kommerell's diverticulum should be resected surgically, regardless of size [7].

Vascular ring can be associated with intracardiac anomalies, such as ventricular septal defect, tetralogy of Fallot, patent ductus arteriosus, and etc. The most common symptom of the vascular ring is known as a noisy breathing (stridor). A large number of patients with a vascular ring show recurrent upper respiratory tract infections and chronic cough, in particular, older children can manifest dysphagia for solid food [2].

Esophagography and CT scan are commonly used for the diagnosis of a vascular ring, and are known to be important diagnostic tools. On occasion, a bronchoscopy is needed to evaluate the degree of tracheal compression. In addition, 2-D echocardiography is needed to find intracardiac anomalies [8].

The left thoracotomy is the recommended modality, as a good surgical approach, for a vascular ring caused by the right aortic arch and left ligamentum arteriosum. If it is intracardiac anomalies, median sternotomy is recommended [2]. The ligamentum arteriosum compressing the trachea and esophagus must be resected completely, and if it is Kommerell's diverticulum, a complete resection of the diverticulum is recommended with an aortic clamp, including the base of the diverticulum with a side-biting vascular clamp. This reduces the risk of recurrent tracheo-esophageal stenosis [7].

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