Case Report

Surgical Translocation of the Circumflex Aortic Arch with Tracheobronchial Compression; A Case Report and Review of the Literature.

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Abstract

Symptomatic Airway compression due to the circumflex aorta is a rare condition. This is a case of 9-year-old boy who had a longstanding history of swallowing problems and was diagnosed as a 22q11.2 deletion syndrome. CT chest images showed a right aortic arch with a circumflex aorta that had looped behind the trachea and esophagus and had a mirror branching with left Subclavian artery arising from the left descending aorta. The airway compression and dysphagia pose a challenge for those patients, as well as the anesthesiologist. We report a case of airway compression in a 9-year-old patient who underwent elective aortic arch reconstruction for the circumflex aortic arch, division, and ligation of the ligamentum arteriosum, and aortoplexy on deep hypothermic circulatory arrest and cardiopulmonary bypass.

Keywords: Airway, Dysphagia, Circumflex, Aorta, tracheomalacia, bronchoscopy

Introduction:

Vascular tracheobronchial compression syndrome could be acquired or congenital, commonly seen with double aortic arch, aberrant subclavian artery, and pulmonary artery sling (1). Kommerell diverticulum is a rare cause of tracheobronchial compression (2). Aneurysms of the aortic arch and descending thoracic aorta can also cause airway compression (3). Chronic compression of trachea can lead to tracheomalacia (4). A Circumflex Aorta is a rare vascular ring caused by a right aortic arch with left ligamentum arteriosum and a descending aorta that crosses posteriorly from the right aortic arch, the left ligamentum and the posterior crossing aorta causing the typical symptoms of noisy breathing, dyspnea on exertion, dysphagia and frequent upper respiratory tract infections. Although ligamentum division would divide the ring, that alone would

not relieve the compression produced from this abnormal vascular anatomy.

We describe the surgical repair in a symptomatic 9-year-old patient who underwent elective Aortic arch reconstruction for the circumflex aortic arch, division, and ligation of the ligamentum arteriosum, and Aortoplexy on Deep Hypothermic circulatory arrest (5).

Case presentation:

A 9-year-old boy with a diagnosis of 22q11.2 deletion syndrome was found to have an aortic arch and left pulmonary stenosis and was referred for further investigation, in addition to genetic and neurology assessment.

A diagnosis of 22q11.2 deletion syndrome was confirmed on FISH testing. From a cardiac standpoint, he was described as active and happy, but his mother noted that he had difficulty in swallowing solid foods. He had no history of syncope or significant episodes of palpitations. Family history included septal defects, coronary heart disease, cleft palate and bone abnormalities of the lower body.

On examination, his weight was 26.8kg, and height was 125.1cm. Heart rate was 91 beats per minute, systemic oxygen saturation was 97% on room air and blood pressure on the right arm was 103/66.

He was alert, comfortable, and not distressed or anemic. His chest was clear bilaterally, and cardiac rhythm was regular. The first heart sound was normally heard while splitting of second heart sound was heard at the base, abdominal examination revealed no organomegaly.

A 12-lead ECG was performed that showed normal sinus rhythm and normal ST-T segments. Echocardiography showed no intracardiac abnormalities and was notable for crossing branch of the pulmonary arteries that have a superior-inferior relationship, that did not appear obstructed but showed a right aortic arch with a probable aberrant left subclavian artery. The arch appeared high and suggested the possibility of a cervical arch. Given the difficulties in resolving his arch anatomy and the potential need to assess his esophagus; Barium Swallow & CT angiography were performed (Figure 1& 2)

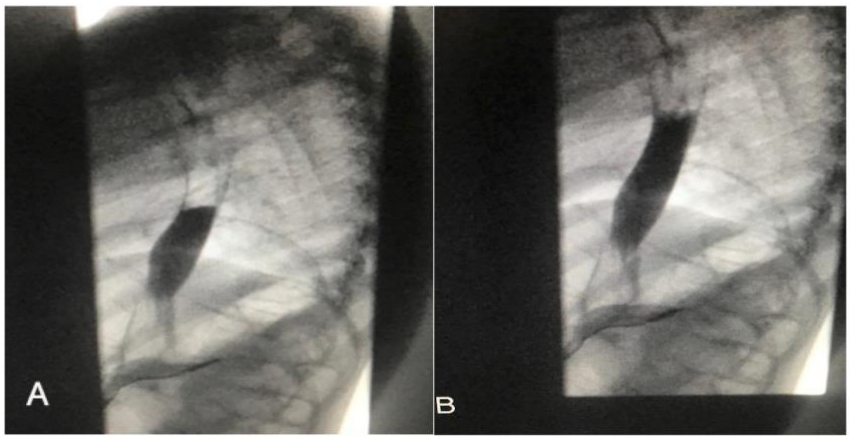

Figure 1. Barium Swallow demonstrating posterior indentation of the esophagus at the thoracic inlet, the site of Kommerell diverticulum

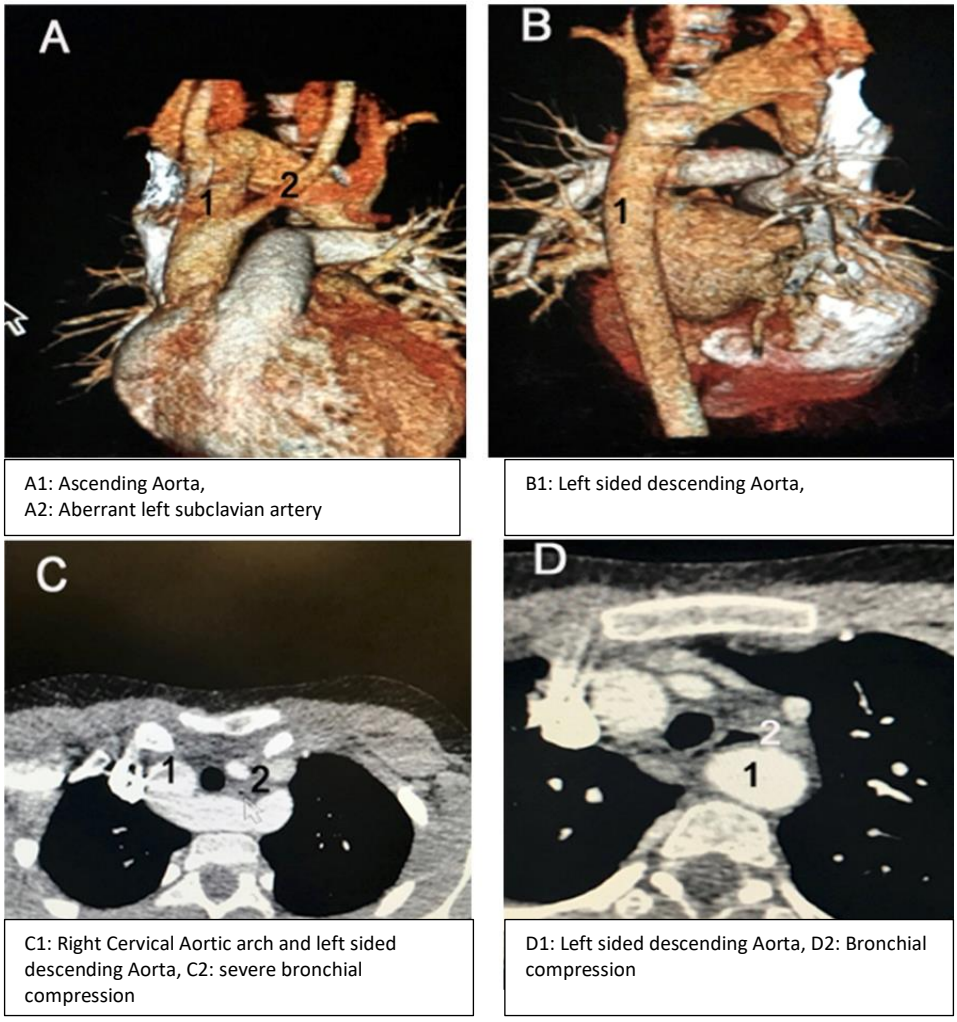

Figure 2. Flinging consistent with a loose vascular ring is seen. There is a right cervical aortic arch and left-sided descending thoracic aorta with an aberrant left subclavian artery and Kommerell diverticulum

Procedure & Operative Findings

The right aortic arch was found to give rise to the left common carotid artery, the right common carotid, and subclavian arteries; it then looped behind the trachea and esophagus in the left side of the chest and descended on the left side of the descending aorta. The left subclavian artery arose from the distal part of the arch on the left side, while the ligamentum arteriosum was also seen at this site.

The aortic uncrossing procedure was performed through a median sternotomy with cardiopulmonary bypass, hypothermia, and a short period of circulatory arrest. The heart was arrested with cardioplegia, head vessels where snared, and deep hypothermic circulatory arrest was established. The aorta was transected distal to the takeoff of the right subclavian artery and the proximal stump oversewn and plexyed to the right side underneath the sternum. The ligamentum arteriosum was doubly ligated and divided. The right and left recurrent laryngeal nerves were identified and preserved. The descending aorta was dissected from its posterior attachments and brought up on the left side of the ascending aorta. Arteriotomy was performed on the side of the ascending aorta adjacent to the left carotid artery and anastomosed to the descending aorta. The circumflex aorta was anastomosed to the ascending aorta with 5.0 prolene suture in an end to side fashion. The circulation was resumed, and the patient warmed and weaned from cardiopulmonary bypass. This procedure relieved both the posterior compression caused by the circumflex aorta and the right-sided compression of the esophagus and the trachea from the right aortic arch. Bronchoscopic examination after circumflex arch repair showed relief of the bronchial compression. The mean circulatory arrest time was 10 minutes. The cardiopulmonary bypass time was 143 minutes, and the cross-clamp time was 30 minutes. There were no postoperative comorbidities or neurological complications, all symptoms were relieved post-surgery. The length of the hospital stay was 10 days.

Discussion:

The majority of patients who had a surgical intervention for a vascular ring have resolution of their symptoms. However, 5% to 10% of those patients develop recurrent symptoms related

either to airway or esophageal compression and may require re-operation**.** Circumflex or encircling of the aortic arch is a rare form of a vascular ring; it is usually associated with severe tracheobronchial compression created by a ligamentum arteriosus or by the retro-esophageal aorta itself (6). The treatment in symptomatic patients consists of surgical ligation and division of the ligamentum arteriosum, the circumflex aorta, and aortoplexy on deep hypothermic circulatory arrest, which usually allows the trachea and esophagus to be freed of compression by this vascular ring and the looped circumflex aorta. Right-sided aortic arch is widely recognized as a cause of tracheobronchial and esophageal obstruction in infants and children, where it forms a complete vascular ring. A circumflex aorta would have both right-sided compression from the right aortic arch as well as posterior compression produced by the circumflex descending aorta**.**The trachea and esophagus are compressed usually from the right aortic arch, the left ligamentum arteriosum and the posterior aorta causing the typical symptoms of noisy breathing, dyspnea on exertion, dysphagia and frequent upper respiratory tract infections. Although ligamentum arteriosum division would divide the ring but would not relieve the compression produced by the abnormal vascular anatomy. The cause of respiratory difficulties in the postoperative period is generally thought to be secondary to bronchomalacia, but we believe that this should be a diagnosis of exclusion, since the symptoms of residual compression may be similar.

References:

1.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;48(3):164-74.
2.Herway ST, Benumof JL, Golts E, Manecke GR. Major positional obstruction of the trachea in a patient with a right-sided aortic arch and Kommerell's diverticulum. J Cardiothorac Vasc Anesth. 2015;29(1):146-8.
3.Koomen E, Schurink GW, Mochtar B, Jacobs MJ, Smets RJ. Repair of thoracic aortic aneurysm associated with tracheal and right mainstem bronchus compression. J Cardiothorac Vasc Anesth. 2007;21(1):88-90.
4.F.G.Pearson. Thoracic Surgery. New York: Churchill Livingstone; 2002. 272-6 p.
5.Backer CL, Mavroudis C, Rigsby CK, Holinger LD. Trends in vascular ring surgery. J Thorac Cardiovasc Surg. 2005;129(6):1339-47.
6.Bruniaux J, Villain E, Sidi D, Almange C, Kachaner J, Lecronier G, et al. [Severe tracheo-bronchial compression caused by a circumflex aorta. Apropos of 3 cases treated by aortic uncrossing]. Arch Mal Coeur Vaiss. 1984;77(5):557-63.