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Right Aortic Arch, Kommerell Diverticulum, and Symptomatic Retro-Esophageal Vascular Ring in an Adult (Case Presentation and Review)

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A 24 year-old Hispanic man presented with progressive retrosternal chest pain, shortness of breath, and dysphagia. Chest X-ray, computed tomography angiography, and magnetic resonance (MRA) studies were compatible with a right aortic arch and a 3.5-cm descending right aortic Kommerell diverticulum. Barium esophagogram and esophagoscopy demonstrated a constricting band crossing the esophagus at the level of the diverticulum. After discussion with other cardiothoracic surgeons, with a combined 300 years of experience, they reported only 1 other possible such case. Kommerell's diverticula are usually repaired in children and rarely are seen in the adult. Kommerell diverticular treatment options include endovascular stenting, cardiopulmonary bypass with resection and grafting, right thoracotomy, and a cervical approach. Because of the patient's findings of a right arch, a Kommerell diverticulum, and a constricting band causing esophageal compression and respiratory distress, it was felt stenting alone would not relieve his symptoms. Therefore, we used a left third posterior intercostal thoracotomy approach to resect the constricting esophageal band connecting the Kommerell diverticulum to the left subclavian artery and to perform the Kommerell

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diverticulectomy. Releasing the esophageal and tracheal compressing band and resection of the diverticulum provided complete symptom relief with elimination of the dysphagia, dyspnea, wheezing, and chest pain, using a single surgical procedure.

Key words: Kommerell diverticulum – Vascular ring – Right aortic arch – Retroesophageal band – Dyspnea – Chest pain in adult – Dysphagia – Surgical resection

Chest pain, shortness of breath, and difficulty swallowing are rather common patient complaints. These complaints may result from either acquired or congenital lesions. More commonly, these problems may be associated in the adult with coronary occlusive or vascular heart disease, gastroesophageal reflux disease (GERD), and chronic pulmonary disease (COPD). Congenital causes of respiratory, cardiovascular, or gastrointestinal complaints usually present in the infant or young child.[1]

The usual causes of respiratory distress in a young individual are pulmonary in nature. These symptoms may also be associated with esophageal abnormalities, especially in the infant. On occasion, vascular ring abnormalities may create both breathing and swallowing difficulties during childhood.

Major congenital vascular obstructive processes with breathing and swallowing difficulties are rarely seen in the adult. These symptoms are even more uncommon in association with a right-sided arch and Kommerell's diverticulum, especially in an adult. We saw a 24-year-old man with severe dysphagia, difficulty breathing, wheezing, and severe substernal chest pain due to a right-sided aortic arch with a concomitant vascular ring and a Kommerell's diverticulum, which is a very rare occurrence and combination. The therapeutic approach considerations for resection of the diverticulum and release of the band were paramount to a single procedure's success compared with other procedural options.

Case Presentation

This 24-year-old Hispanic man was seen with complaints of progressive retrosternal chest pain associated with shortness of breath while at work. In addition, he had a recent onset of progressive dysphagia and wheezing, but continued to work at his job. He had no other medical history, and physical examination by his personal physician was normal. A chest X-ray demonstrated a right-sided aortic arch and no other abnormalities.

Barium esophagogram demonstrated a transverse constriction or narrowing of the upper esophagus above the level of the carina. The computerized tomographic angiogram delineated a descending right thoracic aorta crossing over to the left of the spine at the level of the diaphragm. In addition, a diverticulum of the descending right thoracic aorta was noted projecting to the left with compression of the esophagus (Fig.1). The left carotid and subclavian arteries originated from the right aortic arch anteriorly. The left subclavian artery progressed posteriorly to the left to attach to an esophageal compressing band arising from the Kommerell diverticulum. Magnetic resonance angiography confirmed the findings. Esophagoscopy demonstrated no esophageal irritation or ulceration. Echocardiography and cardiology consultation demonstrated no cardiac, ductus, or ligamentum arteriosum defect.

Surgery

Following preoperative discussion regarding the therapeutic options, risks, and informed consent, a left third intercostal posteriolateral thoracotomy

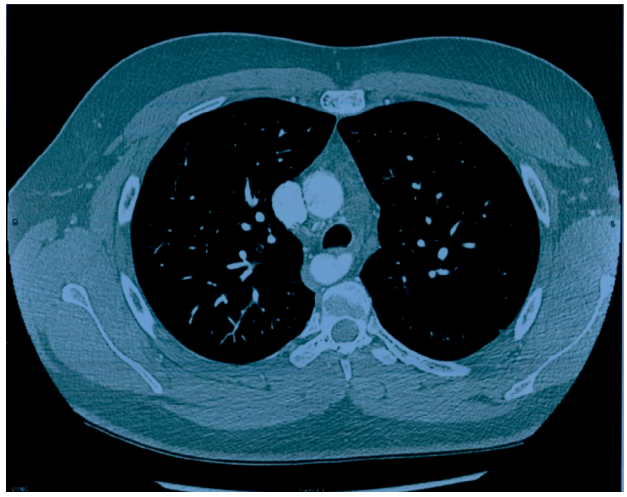

Fig. 1 Computed tomographic angiogram demonstrating the right aortic arch, the right descending thoracic aorta, and the Kommerell diverticulum.

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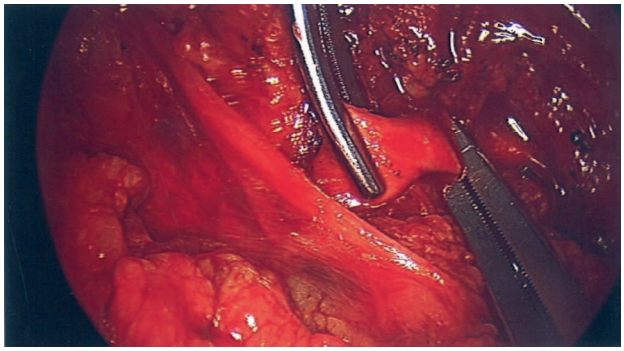

Fig. 2 Medial and lateral vascular clamps applied to the fibrous band (vascular ring) extending from Kommerell diverticulum to the left subclavian artery crossing the trachea and esophagus.

using a double lumen endotracheal intubation single lung ventilation provided excellent exposure directly over the area of pathology. A nasogastric tube was placed in the esophagus during the surgical procedure to enhance esophageal definition. Proximal to the constricting band, the esophagus was markedly distended and thickened. Distal to the constricting band, the esophagus was small and collapsed.

After collapse of the left lung, the posterior mediastinum was opened, and a 1-cm-wide × 3-cm-long fibrous constricting band taking origin from the Kommerell diverticulum and crossing the esophagus to attach to the left subclavian artery was dissected free (Fig. 2). It was clamped medially and laterally and divided, and each end was sutured with 2 layers of running 4-0 Prolene suture to release the esophageal constriction and the tracheal compression and to provide excellent exposure of the diverticulum.

The 3.5-cm aortic (Kommerell) diverticulum was then dissected down to its aortic origin, and a partial occluding vascular clamp was applied to the diverticular base. The diverticulum was resected, and the aortic defect was closed with interrupted prolene sutures using felt pledgets. Postoperatively, his chest pain, wheezing, and shortness of breath were all relieved, and he was able to eat without any dysphagia and was discharged home symptom free a few days later.

Discussion

A right-sided aortic arch and a descending right thoracic aorta are uncommon. Less common is an associated vascular ring and even more rare is an associated Kommerell diverticulum, especially in the adult.[1,2] Early in life, this patient had no difficulty eating, no chest pain, no wheezing, and no shortness of breath. However, as an adult at the age of 24 years, he developed progressive symptomatology that affected multiple systems, including the respiratory system (wheezing and shortness of breath) and the gastrointestinal system (difficulty with passage of food), as well as a rare congenital vascular aneurysm. In addition, he complained of chest pain due to the constriction. Noninvasive X-ray and magnetic resonance testing established the diagnosis.

When considering the various therapeutic options, multiple choices were reviewed to relieve him of his congenital symptomatic anatomic problems. Options available included the placement of a percutaneous transfemoral aortic stent (thoracic aortic endostent) to cover the aneurysm and exclude the lumen. This, however, would not correct the tracheal and esophageal compressive symptoms and allow him to breath and eat without difficulty. Therapeutic options presented in the literature include the following: a right thoracotomy, an endovascular stent, a cervical approach, utilization of cardiopulmonary bypass, and the value of resecting or not resecting the diverticulum and/or the ligament (fibrous band) and the use of vascular staples thorascopically.[2,3]

The aneurysm was not huge and was limited to the descending right-sided aorta. Neither cardiopulmonary bypass nor a 2-step procedure using an endostent and then a thoracotomy was felt to be required. The constricting band required a thoracotomy to be resected and to relieve the gastrointestinal and respiratory complaints. Simultaneous resection of the aneurysm and resection of the constricting band were felt to be appropriate. In other patients, without the constricting symptomatic band, an endovascular procedure without a thoracotomy may be the best option.

Vascular rings are frequently described in medical school and seen occasionally in the infant or small child. Adults with these symptomatic anomalous cardiac and vascular abnormalities are rare. In our experience, the left-sided arch with a retroesophageal right subclavian artery has been the most common adult vascular ring anomaly requiring correction.[4] The adult right aortic arch with an esophageal obstructing band is very unusual to rare.[5] Symptomatology in these patients may also include right subclavian artery dissection, syncope, and acute aortic dissection.[6–8] Cina *et al* developed a classification for the various arch, subclavian, and

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Kommerell diverticulum anomalies.[9] The demonstration of the right arch with the right subclavian artery origin in the normal location in this patient is uncommon to rare.[2] A further concern in this patient was whether he may have a retroesophageal left subclavian artery requiring bypass, but this was not the case. The left subclavian and the left carotid arteries arose anteriorly from the right-sided aorta as a common trunk. In addition, this individual did have a 3.5-cm aortic diverticulum and an aortic diameter of less than 2.5 cm. In most patients with a Kommerell diverticulum, the aorta and the diverticulum are of equal or approximately equal diameter.

Multiple approaches have been used when complicating circumstances exist, such as a diffuse thoracic aneurysm, a giant Kommerell diverticulum, dissection of the aorta, or aortic valvular regurgitation.[6,8,10–12] Necessary corrective procedures may be performed using a single stage, a multiple stage, or endovascular or cardiopulmonary bypass techniques. Gao *et al* performed endovascular grafting and amplatzer vascular plugging of the left subclavian artery (LSA) with the intent to await symptoms prospectively to decide whether a LSA graft will be necessary.[13] We felt an endovascular approach without relief of the constriction would not be appropriate therapy. Fortunately, we were able to follow the concept to "go through the side opposite the arch" and use a single left thoracotomy for resection and alleviation of the patient's multiple symptoms.

Future Perspective

This patient presents the need for a physician to listen to and study all of a patient's symptomatology and needs. When one diagnoses a problem, one should be certain the diagnosis and options for therapy match the presenting complaints by the patient. Thus, a less invasive procedure may not adequately resolve the symptomatology complex encountered and thus may not be the best therapeutic alternative. In situations such as herein presented, treatment of the entire symptom complex requires consideration of each symptom and the structural cause. Correction of 1 defect (the aneurysm) would not have been curative of the entire symptom complex (Kommerell compression and band constriction) and would thus require future invasive and possibly more complex intervention. Either option performed alone—stenting the diverticulum or division of the band—would not accomplish the necessary therapeutic result. Thus, the thoracotomy necessity and the need in future patients to consider the symptom complex and the most appropriate therapeutic approach.

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