Case reports

Surgical treatment of an aneurysm originating from a Kommerell's diverticulum in the right-sided aortic arch with retroesophageal component

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We report the case of a 41-year-old man presenting with a large Kommerell's diverticulum aneurysm in the right-sided aortic arch with retroesophageal component and moderate thoracic aortic dilation. Surgical treatment was performed through left thoracotomy and consisted of aneurysmectomy, closure of the distal aortic arch defect and aorta-left subclavian artery bypass. After 2 years computed tomography showed no modifications in the thoracic aortic morphology and the patency of the graft to the subclavian artery.

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Introduction

In aortic arch anomalies, the persistence of remnant or a stump of the left fourth arch constitutes the arterial diverticulum described by Kommerell[1] in 1936. The development of the diverticulum is dependent on the ductal flow entity, being absent in low ductal flow conditions (e.g. tetralogy of Fallot)[2]. Kommerell's diverticulum (KD) is mostly frequent in right aortic arch with a retroesophageal component[3], classified as type II right aortic arch[4] or type 3 right aortic arch[5] (Table I). This arch abnormality represents the second most common condition of complete vascular rings when the *ligamentum arteriosum* connects the left subclavian artery, or a KD, to the pulmonary artery[6]. Vascular rings are congenital anomalies in which the aortic arch and its branches completely or incompletely encircle the esophagus and trachea and may produce compression. The vascular ring of type II right aortic arch often proves to be loose, with no or mild compression on the mediastinal organs.

Right-sided aortic arch occurs only in 0.05% in the general population and the conditions requiring surgical correction are even more rare.

Type II right aortic arch with KD may be asymptomatic all life long. Tracheal and esophageal compression of this anomaly usually develops in infancy, often within the first month of life, when severe obstructed airway symptoms generally constitute the indication for intervention. Clinical manifestations of borderline severity usually disappear with growth, but signs and symptoms can appear when KD and/or thoracic aorta became aneurysmal. These patients may be prone to aortic dissection in the right aortic arch. The most frequent clinical manifestations reported in adulthood are related to the compression of the esophagus and/or the superior airways. Symptoms are dysphagia, dyspnea, cough, chest pain, and asthma[7-9]. The operation is performed either to resolve the mediastinal compression or to resect a right aortic arch and/or a Kommerell's diverticulum aneurysm (KDA).

KDA is a very rare but clinically relevant condition because of mortality associated with rupture and dissection, morbidity caused by compression, and complexity of treatment.

In 1985 Austin and Wolfe[10] reviewed 32 cases of KDA and reported an operative mortality of 16.6% for elective surgery in 18 patients, showing a complex and high-risk surgery. It is remarkable that 19% of the patients presented with rupture and all of them died. Cina et al.[11] have recently re-

Table I. Classification of right-sided aortic arch.

Edwards[4], 1948	Shuford and Sybers[5], 1974
Type I With mirror-image branching of major arteries	Type 1 Embryonic left arch interrupted distal to the ductus arteriosus. Mirror-image branching of major arteries
Type II With aberrant subclavian artery	Type 2 Embryonic left arch interrupted proximal to the ductus arteriosus. Mirror-image branching of major arteries and retroesophageal ligamentum arteriosum
Type III With isolation of the subclavian artery. The subclavian artery is connected to the pulmonary artery through the ductus arteriosus. The ductus arteriosus may be on the left, on the right or bilateral	Type 3 Embryonic left arch interrupted between the left subclavian and the left common carotid arteries. Retroesophageal left subclavian artery and ligamentum arteriosum
	Type 4 Embryonic left arch interrupted between the left common carotid artery and the right arch

viewed 29 cases of KD and thoracic aortic aneurysms with right-sided aortic arch, reporting an operative mortality of 8.3% for elective surgery and 18% for aneurysms associated with dissection.

Case report

A 41-year-old man symptomatic for angina-like chest pain was admitted to our community hospital. Acute coronary syndrome was excluded with ECG and troponin measurement. Chest roentgenogram showed enlargement of the aortic knob and subsequent computed tomography demonstrated a 5.5-cm spherical aneurysm originating from a KD in the right-sided aortic arch with retroesophageal component (Edwards type II). The KDA occupied the upper anterior left mediastinum and communicated with the distal aortic arch through the retroesophageal space. Distal aortic arch and the descending thoracic aorta were moderately and uniformly dilated (main diameter 41.5 mm). The aortic distal arch occupied the midline of the upper posterior mediastinal space (Fig. 1). The thoracic descending aorta was right-sided. An aberrant left subclavian artery originated from the KDA. Diagnosis was confirmed by angiography (Fig. 2).

The patient was scheduled for surgical intervention to prevent rupture of the KDA. Signs and symptoms of compression were absent. The aim of surgery was aneurysm resection and left arm revascularization with an ascending aorta-left subclavian artery bypass.

Left anterolateral thoracotomy in the third intercostal space was extended toward the posterior axillary line to obtain a better vision of the posterior mediastinum. The aneurysmal sac was largely adhesive to the wall of the distal arch. Resection with tangential aortic cross-clamping, without cardiopulmonary bypass (CPB), was not performed because of the absence of a well-delimited aneurysm neck, the risk related to the extensive dissection on fragile and adhesive tissues and the poor surgical view of the right-sided aorta. CPB was instituted with right atrial and left femoral arterial cannulation and a brief period of hypothermic cardiocirculatory arrest (22°C) was performed. The aneurysm incision showed a thin wall and no endoluminal thrombosis. Low flow CPB was re-instituted to minimize air embolism. The aortic origin of the sac, a circular hole of 5 mm in diameter, was closed with a direct suture, reinforced with a "capitonnage" of the aneurysm wall because of tissue fragility. *Ligamentum arteriosum* was absent and there was no evidence of complete vascular ring.

The ascending aorta-subclavian bypass graft was performed with 10-mm tubular Dacron prostheses, receiving an "L-shaped" orientation to avoid any compression on the upper mediastinal organs.

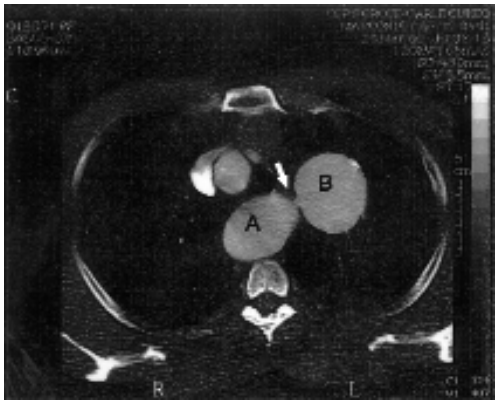
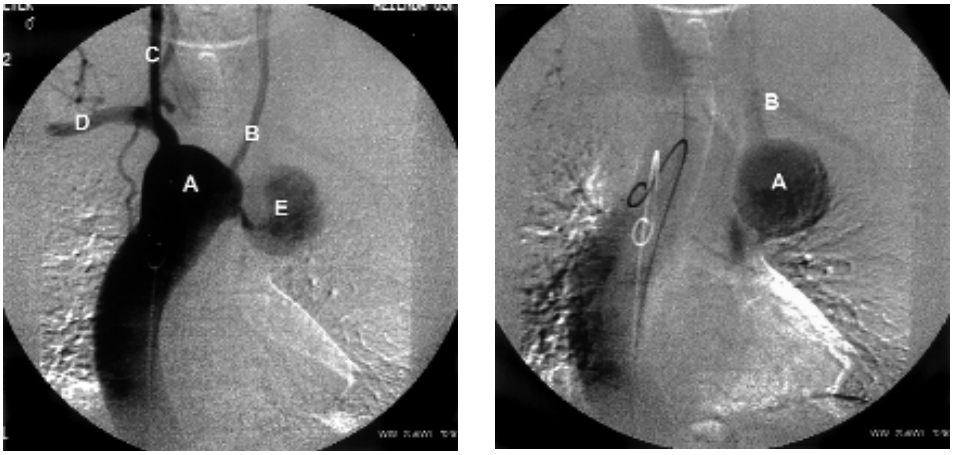


Figure 1. *Preoperative computed tomography axial scan. A = distal aortic arch; B = Kommerell's diverticulum aneurysm. The* white arrow *shows the communication between the distal aortic arch and the Kommerell's diverticulum aneurysm. Distal aortic arch kinks rightward, being the thoracic descending aorta right-sided.*



A B

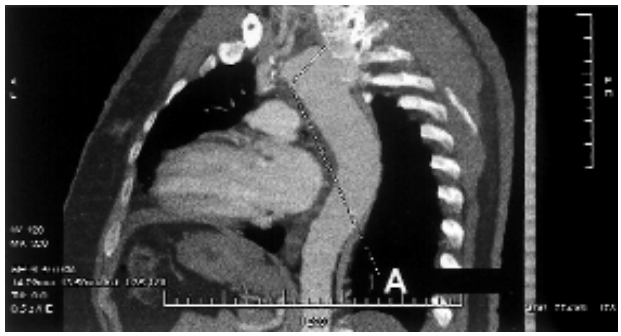
Figure 2. *Preoperative transfemoral angiography of the ascending aorta and aortic arch.* A: *early phase with partial opacification of the Kommerell's diverticulum aneurysm. A = ascending aorta; B = left common carotid artery; C = right common carotid artery; D = right subclavian artery; E = Kommerell's diverticulum aneurysm.* B: *late phase with complete opacification of the Kommerell's diverticulum aneurysm. A = Kommerell's diverticulum aneurysm; B = left subclavian artery.*

The recovery was uneventful and the patient was discharged 8 days later. Seven-day computed tomography control demonstrated a complete resection of the aneurysmal lesion and patency of the aorta-subclavian bypass. Histological examination of a sample tissue from the aortic defect fringe showed mucoid degeneration. At 2-year follow-up the patient was asymptomatic. Computed tomography scan showed unvaried dimensions of the thoracic aorta and patency of the aorta-subclavian bypass (Fig. 3).

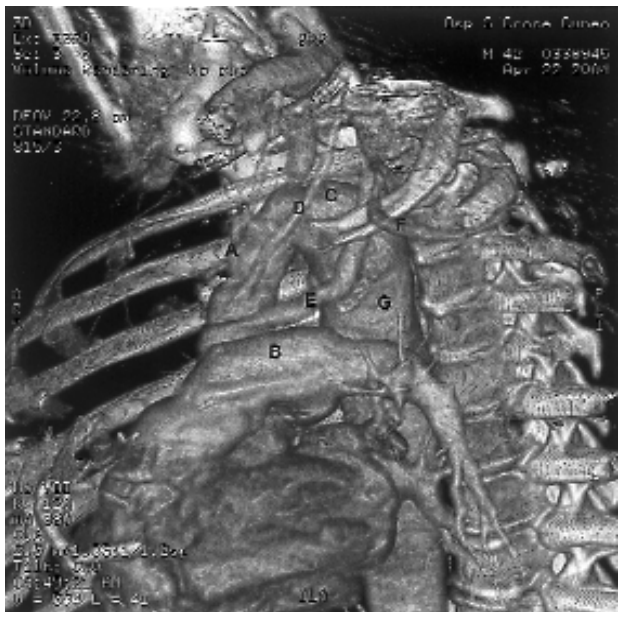
Discussion

In childhood right aortic arch with retroesophageal component and KD is generally treated by means of left thoracotomy[12,13]. This approach provides a good exposure for the division of the vascular ring and the resection of the KD and allows a simultaneous revascularization of the left arm with left subclavian artery to the left common carotid artery transfer[14].

In adulthood, in the presence of KDA and/or thoracic aortic aneurysm, right thoracotomy is generally used, because it provides a good surgical exposure of the ascending, transverse arch and descending thoracic aorta[11]. In these cases surgical intervention consists of resection of the aneurysmal sac, interruption of the vascular ring, and revascularization of the left arm. When the aneurysm originates from a KD and extends to the descending aorta, a tube graft is used to replace the aneurysmal aorta and the left subclavian artery is attached to the tubular graft with an interposition graft[15]. Alternatively a left subclavian-left common carotid artery transfer can be performed through a second left cervical incision[11,15]. Cina et al.[11] indicate endoaneurysmorraphy through right thoracotomy as the ideal treatment for small KDA with normal descending thoracic aorta.



A



B

Figure 3. *Two-year postoperative computed tomography.* A: *maximum intensity projection showing distal aortic arch. A = aortic arch diameter at the site of aneurysmectomy (41.9 mm).* B: *volume rendering reconstruction. A = ascending aorta; B = pulmonary artery; C = aortic arch; D = left common carotid artery; E = aorta-subclavian bypass; F = left subclavian artery; G = descending thoracic aorta.*

Jung et al.[16] recommend right thoracotomy in the presence of a large KD. Mossad et al.[17] report a left thoracotomy approach in a review of a series of KD, but describe clearly only one case of a small diverticulum (2 cm in diameter), associated with hypoplastic right aortic arch, resected without CPB. Midline sternotomy is rarely used and may fail to give a good surgical exposure, requiring concomitant thoracotomy[18]. Bilateral thoracotomy is rarely used[19].

Our report describes the surgical treatment of a large KDA in Edwards type II right aortic arch, associated with moderate dilation of the thoracic aorta, through a left thoracotomy approach.

Our procedure was directed to KDA treatment and left arm revascularization in a single surgical stage. In our experience the possibility of applying an "off-pump" procedure safely seems to depend on the diameter of the aneurysm and the good exposure of the thoracic aorta at the origin of the sac. A left approach allowed us to perform an easy graft interposition between the aorta and the left subclavian artery in a single surgical session. Although subclavian-to-carotid transposition before intrathoracic repair via right thoracotomy is suggested as an ideal technique for reconstruction of this artery[11], we believe that the possibility of performing a single surgical approach, of avoiding carotid artery manipulation and restoring a more physiological arterial flow constitute an advantage in favor of left thoracotomy. In our strategy the moderate dilation did not justify a more technically demanding and potentially hazardous procedure on the descending thoracic aorta. However, the persistence of dilated aortic arch and descending aorta can represent a prognostic problem because of the potential aneurysmal evolution and should be controlled in mid- and long-term follow-up. The finding of a mucoid degeneration of the arterial wall, which potentially could predispose to aortic dilation, seems to reinforce this consideration, although the persistence of fragile tissue in the site of the aortic suture has not been correlated with the risk of complications in previous reports.

Our experience suggests that a single session resection and aortic defect closure, associated with aorto-subclavian bypass, constitutes an effective choice for the treatment of this complex anomaly, and complications related to mild aortic dilation and arterial wall fragility were absent at 2-year follow-up.

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