Case report - Congenital

Asymptomatic pseudo-aneurysm of the aortic arch in a patient with aberrant right subclavian artery. A complication of Kommerell’s diverticulum?

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Abstract

Kommerell’s diverticulum is an aortic arch deformity associated with an aberrant subclavian artery. Symptoms related to compression of adjacent structures, dilatation of the aortic diverticulum or accelerated atherosclerosis leading to increased risks of dissection and rupture represent the indications for surgical treatment. Several surgical strategies have been used for the management of this congenital abnormality. We present the case of a 56-year-old male with a pseudo-aneurysm of a left aortic arch adjacent to a Kommerell’s diverticulum at the orifice of a left subclavian artery. The patient also presented an aberrant right subclavian artery originating from the posterior wall of the ecstatic take-off of the left subclavian artery. Our surgical strategy was limited to the resection of the aneurysm without any manipulation of the aortic diverticulum and aberrant right subclavian artery, as the wall of both aorta adjacent to the saccular aneurysm and left subclavian artery was normal, the Kommerell’s diverticulum was small and the patient was asymptomatic.

Keywords: Kommerell; Aortic arch; Pseudo-aneurysm; Congenital; Aberrant right subclavian artery

1. Introduction

Kommerell’s diverticulum (KD) is a rare congenital abnormality of the aortic arch occurring in association with an aberrant right subclavian artery (ARSA) in a patient with left aortic arch (LAA), or with an aberrant left subclavian artery (ALSA) in a patient with right aortic arch (RAA) [1]. Burckhard F. Kommerell described a diverticulum of the thoracic aorta at the origin of the ARSA, as a remainder of the primitive right dorsal aorta [2]. In the literature, KD is defined as an aneurysmal diverticulum of the proximal descending thoracic aorta or aortic arch and/or aneurysmal orifice of an aberrant subclavian vessel. These variations together with the rareness of this entity are the reasons for having not yet established either the indications or the strategy for the surgical treatment.

2. Case presentation

A 56-year-old male was referred to our unit due to an incidental finding in the chest X-ray of a tumor-like lesion of the upper mediastinum. The CT-scan revealed a saccular aneurysm of the distal aortic arch. A three-dimensional CT angiography confirmed the diagnosis of a 47-mm diameter saccular aneurysm adjacent to the Kommerell’s diverticulum (Fig. 1a). The descending thoracic aorta had normal diameter. Digital subtraction angiography showed a normal diameter ascending aorta and a left aortic arch with three branches; right common carotid, left common carotid and an ecstatic left subclavian artery. An aberrant right subclavian artery originated from the posterior wall of the dilated left subclavian artery (Fig. 1b).

A left posterolateral thoracotomy was performed through the 4th left intercostal space and the aortic arch, left subclavian artery and descending thoracic aorta were exposed and vascular tapes were placed around them (Fig. 2a). The aneurysm was dissected from the left upper lobe with caution, as it was extremely thin walled (pseudo-aneurysm). Under normothermic partial left femoro-femoral bypass the above vascular areas were clamped and the pseudo-aneurysm was opened and resected. The arterial wall of the neighboring structures did not show any signs of degeneration and since the patient was asymptomatic we decided to put a Dacron patch in the aortic arch at the area of the neck of the pseudo-aneurysm (Fig. 2b). The patient tolerated the procedure well. His postoperative course was uneventful. He was discharged the 7th postoperative day and remains asymptomatic with no signs of enlargement of his Kommerell’s diverticulum 6 months postoperatively.
The aberrant right subclavian artery and white arrow denotes the pseudo-subclavian with the Kommerell' diverticulum at the origin, arrowhead shows the aberrant right subclavian artery and white arrow denotes the pseudo-aneurysm just near the Kommerell's diverticulum. Asterisk denotes right common carotid artery, double asterisk denotes origin of left subclavian with the Kommerell' diverticulum at the origin, black arrow shows the descending thoracic aorta, and arrowhead denotes the ascending aorta.

3. Discussion

Burckhard F. Kommerell described ‘his’ aortic diverticulum in 1936 in a patient with LAAR and ARSA [2]. In the most common type of KD, the right subclavian artery arises as the last branch of the aortic arch and travels by the proximal descending aorta to the right arm, passing behind the esophagus. It is considered as an anomaly of the 4th aortic arch and results from the regression of the right aortic arch between the right carotid and right subclavian arteries.

KD can exist with other vascular anomalies, such as RAA with ALSA, and LAA with right descending thoracic aorta and right ductus arteriosus or ligamentum arteriosum [3].

The majority of patients with this congenital abnormality remain asymptomatic, like our patient. Symptoms may occur due to dilatation of the diverticulum, development of atherosclerosis and increase in rigidity or tortuosity of the aberrant subclavian vessel or as a result of compression of the esophagus or trachea. This might lead to dysphagia, dyspnea, stridor, wheezing, cough, recurrent pneumonia, obstructive emphysema, or chest pain [4]. Catastrophic complications, such as dissection and or rupture are not rare. In the review by Austin and Wolfe, 19% of their patients experienced rupture and all of them died [5]. It may be that the congenitally diseased areas present an accelerated degeneration of the aortic wall resulting in the formation of a pseudo-aneurysm (thin wall) with a high possibility of rupture.

Different surgical approaches have been proposed for the enlarged KD (diameter ≥ 50 mm) or in symptomatic patients. Key components are the excision of the diverticulum and the reconstruction of the aberrant subclavian artery. One or two stage procedures, such as total arch replacement, descending thoracic aorta replacement with subclavian–carotid artery transposition, stent grafting with extra-anatomic bypass, carotid to aberrant subclavian artery bypass and ligation of the subclavian artery proximal to the origin of the vertebral artery, are some of the proposed surgical techniques. Most of these techniques require thoracotomy, left-heart bypass, hypothermic cardiopulmonary bypass, and probably circulatory arrest [6–8].

We proceeded with a more conservative approach, the resection of the pseudo-aneurysm and patching of the defect in the arch wall, as this was the main threat to the patient’s life and the adjacent aortic wall was evaluated as normal. The same is true for the left subclavian artery despite the ecstatic appearance. The aberrant right subclavian artery originated from the posterior wall of this ectatic vessel and the proximity of this subclavian trunk to the left common carotid artery would necessitate hypothermic arrest for the surgical correction. This not only would have added unnecessary perioperative risk but would have required a postoperative revascularization procedure for the right arm due to ligation of the ARSA.

Kommerell’s diverticulum is a rare congenital abnormality. The variability of the clinical and angiographic picture of this vascular anomaly poses a challenge for optimal surgical management, with a wide spectrum of one or two stage procedures. In the case of a pseudo-aneurysm associated with KD, an earlier surgery is justified since the risk of rupture is greater. A more conservative approach, resection and patching, may still be useful for such patients.

References

We appreciate the recent report by Dr. Panagiotou and coworkers describing the case of a 56-year-old male with a pseudo-aneurysm of a left aortic arch adjacent to a Kommerrell’s diverticulum at the orifice of the left subclavian artery [1].

We would like to credit Professor Dr. Burckhard F. Kommerell (1901–1990) for his description of an aortic diverticulum associated with an aberrant right subclavian artery [2]. As chief of the section of radiology at the Charite University Clinic in Berlin since 1934, he published the first report in 1936 with the following description:

‘Thus far an aberrant course of the right subclavian artery has not been reported in a living patient. In these circumstances it seems appropriate to publish an observation that I have made while performing a radiologic examination of the stomach. A 65-year-old man was examined because of a presumed diagnosis of stomach cancer. The sagittal view of a barium swallow examination demonstrated a delay in the passage of contrast medium at the level of the aortic knob. The aortic knob was left-sided and did not show any abnormality except for a small calcium deposit in its wall. Repositioning of the patient in a more oblique direction demonstrated that the esophagus was pushed forward at the level of the aortic knob. To the left and behind the esophagus there was a mass, that, because of its pulsatile character, was interpreted as being a vessel. At this location the esophagus was compressed […]. The trachea and the other thoracic organs were all normal, as were the clinical findings. This radiologic finding can only be interpreted as being an aberrant origin of the right subclavian artery. However, the pulsating mass behind the esophagus does not consist of the right subclavian artery itself, because the calibre of this vessel is much smaller. Much more likely this mass consists of an aortic diverticulum, from which the right subclavian artery originates.’

Kommerell described a patient who had a left aortic arch and an aberrant right subclavian artery. In this vascular anomaly, the right subclavian artery arises as the last branch of the aortic arch and courses from the proximal descending aorta to the right arm, passing behind the esophagus. This anomaly of the 4th aortic arch results from regression of the right aortic arch between the right carotid and right subclavian arteries. Kommerrell pointed out that a diverticulum at the origin of the aberrant right subclavian artery is a remainder of the primitive right dorsal aorta.

We encountered a 55-year-old male suffering acute thoracic pain, vertigo and hypertension with a rupture of a Kommerrell’s diverticulum in a right aortic arch in aortic dissection Stanford type B [3]. This patient underwent emergency distal aortic arch replacement and subsequent bilateral subclavian artery bypass grafting in a single procedure.

References

