

Kommerell Diverticulum in Adults: Evaluation of Routine CT Examinations

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Objective: To evaluate of Kommerell diverticulum (KD) in adults during routine CT examinations.

Material and methods: Eighty-seven cases of left aortic arch with aberrant right subclavian artery (LAARS) and 28 cases of right aortic arch with aberrant left subclavian artery (RAALS) were found on routine CT examinations using 64-128 multidetector CT. We assessed the incidence of KD and measured the size of KD, and compared the results between both groups. We evaluated associated congenital and acquired cardiovascular diseases on both groups. **Results:** The incidence of KD in each group was as follows: RAALS 100 % and LAARS 48.3 % and incidence of KD in RAALS group was significant higher than in LAARS group. The mean KD size was as follows: RAALS 32.8 mm and LAARS 17.8 mm and, the size of KD in RAALS group was larger than that in LAARS group. Two cases were associated with congenital heart disease. Several cases were associated with acquired aortic diseases including aortic aneurysm, dissection, severe atherosclerosis and aortitis.

Conclusion: KD was common among adults with an ASA and some adults were associated with aortic diseases.

Key words: Kommerell diverticulum, CT, adults, aberrant subclavian artery

INTRODUCTION

An aortic diverticulum (AD) characterized by a bulb-like dilation of the proximal descending aorta is called a Kommerell diverticulum, which is often found in a right aortic arch with aberrant left subclavian artery (RAALS) [1-4]. An AD with or without an associated aberrant subclavian artery (ASA), is an uncommon arch anomaly, however AD of ASA was called as Kommerell diverticulum (KD) [1-9]. KD was defined by Becker *et al.* as a widening of the base of the subclavian artery to >1.5 times the size of the distal subclavian artery [7]. The KD or ASA can compress the esophagus or trachea, causing dysphagia, stridor, or asthma [3-5]. In the absence of symptoms, AD has been considered a benign anatomic variant [3, 5, 9]. However, reports have described spontaneous rupture, dissection, and death associated with AD [3, 5, 7, 9]. Approximately 20-60% of individuals with an ASA are affected by KD [3, 7, 8]. Using angiographic examinations, Fisher *et al.* reported that the incidence of KD among patients with a left aortic with aberrant right subclavian artery (LAARS) was 37% (7/19), and the incidence of AD among patients with right aortic with aberrant left subclavian artery (RAALS) was 100% (3/3) [6]. However, the sample size of that study was small. In our study we hypothesized that KD is not a particularly rare condition in adults with or without associated vascular disease. Therefore, we evaluated the incidence of KD in adults during routine CT examinations.

MATERIALS AND METHODS

This retrospective study was approved by the institutional review board of our hospital; the requirement for informed consent was waived because of the retrospective nature of the study.

From January 2006 to December 2015, 45 adults with a right aortic arch and 87 adults with LAARS were recorded in radiological reports of CT examinations. Seventeen adults of right aortic arch with mirror image were excluded. Our study included RAALS group of 28 cases (19 men, 9 women, mean age = 62.6 ± 18.3 years) and LAALS group of 87 cases (43 men and 44 women, mean age = 65.7 ± 16.4 years) included. All right aortic arch cases had a right-side descending aorta, and all left aortic arch cases had a left-side descending aorta. Eleven cases of RAALS were observed on non-contrast CT, and 17 cases of RAALS were identified through contrast-enhanced CT including 6 CT angiographic examinations. Of the LAARS cases, 21 and 60 patients were identified via non-contrast and contrast-enhanced CT in including 29 CT angiographic examinations, respectively. The indications for CT angiography included the suspicion or ruling out of aortic diseases such as aneurysm, dissection, and atherosclerosis, injury, or pulmonary emboli. CT was performed using a 64-128-slice scanner (Somatom Sensation Cardiac 64, Definition, Definition Flash, Definition Edge, Definition AS+, Siemens AG, Munich, Germany) at a slice thickness of 0.5 mm. The other parameters were as follows: 120kVp, 125-250 mA, and a

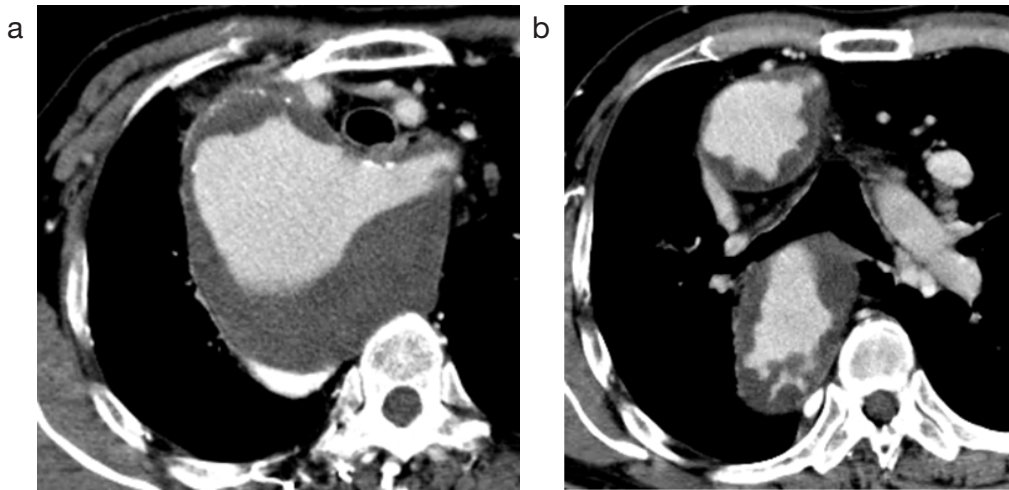


Fig. 1 A large aneurysm of Kommerell diverticulum in a 78-year-old man with a right aortic arch. a. An axial CT image of arterial-phase shows a Kommerell diverticulum aneurysm with a diameter of 88 mm arising from the aberrant left subclavian artery. b. Note the marked atherosclerotic changes in the right ascending and descending aorta.

rotation time of 0.5 s. Conventional contrast-enhanced CT examinations were performed by injecting 2 ml/kg of nonionic contrast material at a rate of 2 ml/s with a 120-s scanning delay. CT angiographic examinations were performed using 100–150 ml of nonionic contrast material injected at a rate of 4 ml/s, and the scanning delay was determined using a bolus tracking method. Two radiologists, each with more than 20 years of experience with CT image interpretation, reviewed the 1–5-mm reconstructed axial CT images on a picture archiving and communication system workstation. Additionally, for cases with CT angiography data, multi-planar reformations and volume-rendering images were used for the evaluations. The incidence of KD was evaluated on both non-enhanced and contrast-enhanced images. The radiologists resolved any assessment disagreements through consensus. If KD was present, the size of the KD was measured. KD was defined as a widening of the base of the subclavian artery to > 1.5 times the size of the distal subclavian artery. According to the measurement procedure defined by Tanaka *et al.*, the maximum distance from the diverticulum wall adjacent to the trachea to the opposite aortic wall was measured [3]. We used the mean KD diameter, calculated from the measurements made by the 2 radiologists, as the measured diameter. Other associated vascular anomalies and aortic diseases were recorded for each case. We used Fisher's test to compare the incidence rates and sizes of KDs among the 2 groups. A P-value < 0.05 was used to define statistical significance. The ages of patients with KD and sizes of KDs were analyzed using a t-test and correlation coefficients. The influence of age (mean age and dispersion) on the presence of KD was analyzed using the median test. The sex-based incidence of KD was analyzed using the chi square test.

RESULTS

A total of 70 KDs (RAALS: 28 and LAARS: 42) were found, and the incidence of KD was 60.9% (70/115). The incidence rates of KD were 100% in the RAALS group and 48.3% in the LAARS group. The incidence of KD was significantly higher

in the RAALS group than in the LAARS group ($P < 0.0027$). The overall mean KD size was 25.1 ± 13.8 mm (RAALS: 32.8 ± 15.2 mm, LAALS: 17.8 ± 13.0 mm). The largest KD, with a diameter of 88 mm, was observed in the RAALS group; the case had severe arteriosclerosis and a diagnosed aortic arch aneurysm with AD (Fig. 1). The largest AD in the LAARS group measured 26 mm in diameter and resulted from arteriosclerosis. No overall correlation was observed between the patients' age and AD size ($t < 1.782$). Similarly, there was no correlation between the presence of AD and the patients' age (chi square = 0.5). Additionally, no significant correlation was observed between the presence of AD and the patients' sex (chi square = 0.96). Patent ductus arteriosus was observed in 1 case of LAARS. Ventricular septal defect was observed in 1 case of RAALS. Four cases with a bi-carotid trunk were observed in the LAARS group (Fig. 2, 3). Two cases with LAARS had a double inferior vena cava. Severe arteriosclerosis was observed in 8 patients. Associated AD aneurysms were observed in 6 cases. Aortic dissection was observed in 2 cases with LAARS (Fig. 2). Aortitis was observed in 1 case with LAARS (Fig. 3). Five patients exhibited AD-related symptoms such as dysphagia, chest pain, and esophageal obstruction and 3 patients had dysphagia lusoria including aortic aneurysm.

DISCUSSION

Congenital anomalies of the aortic arch complex range from asymptomatic normal variations in arch vessel branch patterns to symptomatic vascular rings, stenoses, and arch interruptions [2–6]. The frequency of aortic arch anomalies ranges from 0.5% to 3.0% [4]. KD is a developmental error involving a remnant of the fourth dorsal aortic arch and was named after Dr. Kommerell, a radiologist who first diagnosed this condition in a living individual [1]. KD occurs as an ASA that arises from either the right or left aortic arch to the contralateral side [3]. The ASA is the most common intrathoracic major arterial anomaly, and the reported incidence rates of LAARS and RAALS are 0.7–0.2% and 0.04–0.4%, respectively [3]. Edwards previously

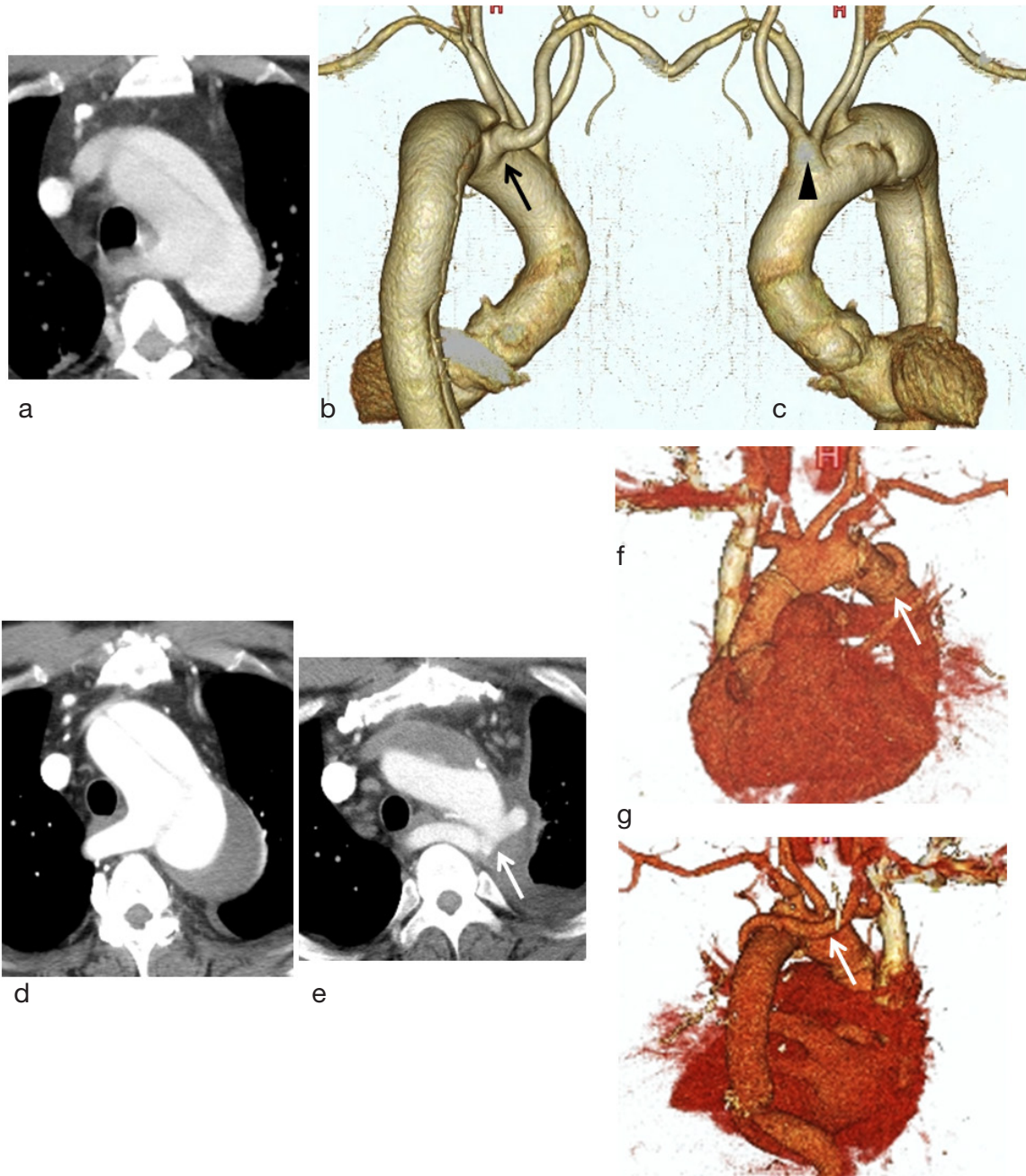


Fig. 2 A Stanford type A aortic dissection in a 39-year-old man with a left aortic arch with aberrant right subclavian artery. a. Axial CT image on the first onset of aortic dissection shows aortic arch dissection including aberrant left subclavian artery. A posterior (b) and anterior (c) views of a 3-dimensional CT image after repair of the ascending aorta show an aberrant right subclavian artery (black arrow), bi-carotid trunk (arrowhead) and remaining aortic dissection of the descending aorta. d. Axial CT image after the first operation shows increasing size of dissecting aneurysm. e-g. CT images after the second operation show graft replacement and reconstruction of the right aberrant subclavian artery (white arrows).

explained the developmental errors and variations of the aortic arch [10]. LAARS results from interruption of dorsal segment of right aortic arch and RAALS results from interruption of dorsal segment of left aortic arch (Fig. 4) [2-4]. Proto *et al.* reported an incidence rate of 60% for the appearance of KD with LAARS as an aortic knob on postero-anterior radiographs [11]. Backer *et al.* reported that the incidence of KD with RAALS was 14.5% in a series of pediatric surgical repair cases [12]. According to a report of adult patients by Fisher *et al.*, the incidence of KD with RAALS, and LAARS was 100% and 37%; however, these authors

evaluated 22 cases via angiography or CT [6]. In our study, the incidence of KD with RAARS and LAARS was 100% and 51.9%; the reported incidence of KD with RAARS in the studies by our group and Fisher *et al.* were identical. The incidence of KD with LAARS was higher in our study than in the study by Fisher *et al.* This difference might be attributable to the sample size and evaluation modality.

KDs with LAARS are often conical, whereas those with RAARS are large and round [3]. The AD size remains unclear in asymptomatic adults. In our study, KDs with RAARS was larger (RAALS: 32.8 mm) than

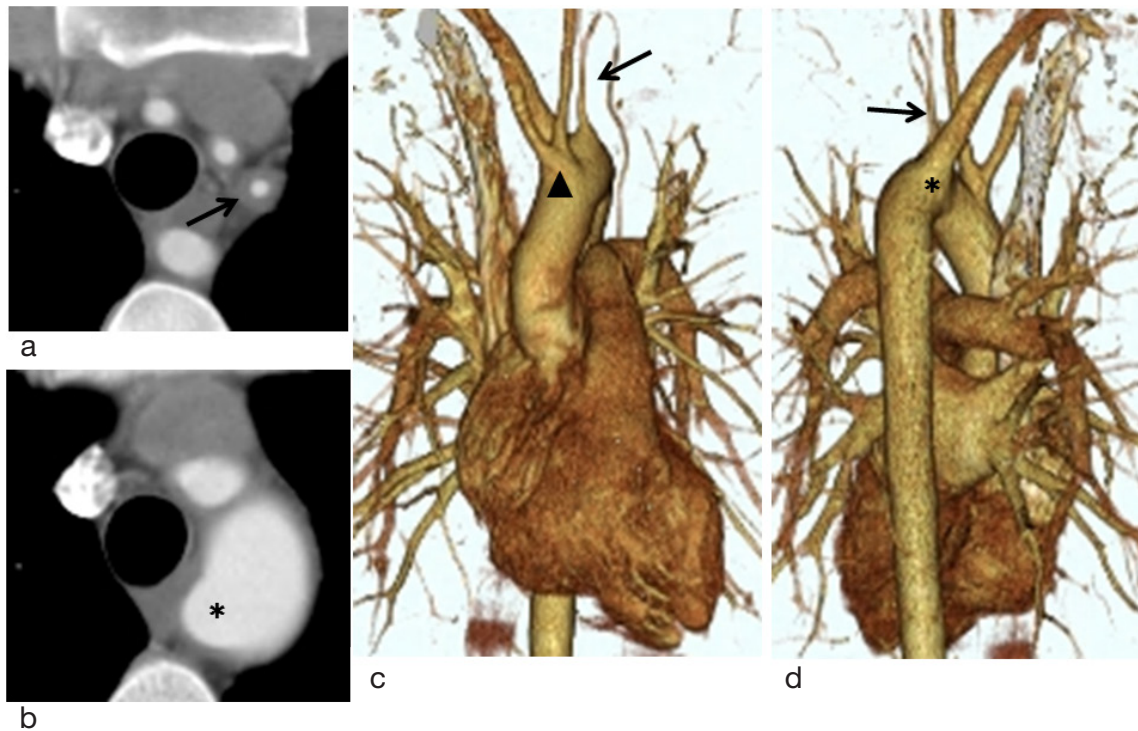


Fig. 3 Aortitis a 28-year-old male

a, b: An axial CT images show a stenosis of the left subclavian artery with wall thickening and Kommerell diverticulum arising from an aberrant left subclavian artery. c. An anterior view of 3-dimensional CT image shows a bi-carotid trunk (arrowhead) and stenotic left subclavian artery. d. A posterior view of the 3D CT image shows a Kommerell diverticulum. Asterisks show Kommerell diverticula and arrows show stenotic left subclavian arteries.

those with LAARS (17.8 mm). Through a literature review following a Medline database search, Tanaka and colleagues identified 212 cases of KD with a mean age of 41.8 ± 26.6 years; 63 patients (30%) were younger than 20 years. The natural history of AD is unknown, given the rarity of this condition [3]. Among patients with KD, the rates of complications such as rupture and/or dissection at the first encounter range from 0% to 50% [3].

The risks of rupture and thromboembolism are high, and the post-rupture mortality rate exceeds 50% not necessarily related to the size of the sac [13]. In our study, the patient with the largest KD aneurysm (88 mm in diameter) could not undergo surgery because of severe pulmonary disease (Fig. 2). According to the literature review by Tanaka *et al.*, the incidence of aortic dissection with AD was 11% [3]. In our study, one patient with LAARS had a Stanford type A and had undergone ascending aortic repair (Fig. 4). Because the size from aortic arch to descending aorta with dissection had been came larger than that of the first operation, the second operation was performed. An aberrant right subclavian was repaired by graft from aortic arch. Bi-carotid trunk is associated with a LAARS commonly [14–16]. On our study 4 cases with bi-carotid trunk were found on LAARS group (Fig. 2, 3). Because bi-carotid trunk can be a risk factor for surgery, the anatomy of ASA, KD and bi-carotid trunk needs to be clearly depicted and described [14]. Given the complexity of surgical treatment for larger lesions, Cina *et al.* advocated treating only aneurysms measuring > 30 mm in diameter [9]. However, in our study, the mean KD size in RAASA was approximately 33 mm. Saccular aneurysms are

believed to carry a significant risk of rupture and can potentially induce an aortic dissection in the KD [17]. Therefore, round and large KDs should be followed up with caution. Also several treatments for KD have been reported, including total diverticulum resection, endovascular repair, and hybrid repair [3, 5–9, 16–21].

Our study had several limitations. First, the sample size was small. Second, the method used to measurement KD size was problematic. For KD, 3D volumetry is superior to 2D measurements. However, 3D data and multiplanar reformatted images were not available for all cases in our study, and therefore we could not use those methods. A large study involving autopsies is needed to determine the true incidence of KD and evaluate of associated vascular disease.

In conclusion, KD was common among adults with a ASA. Our routine CT examinations detected a mean KD size in adults of approximately 25 mm. In the absence of symptoms, KD in adult patients is considered a benign condition by most radiologists. According to some surgeons, adults with KD should be observed more carefully because KD was occasionally associated with acquired aortic diseases.

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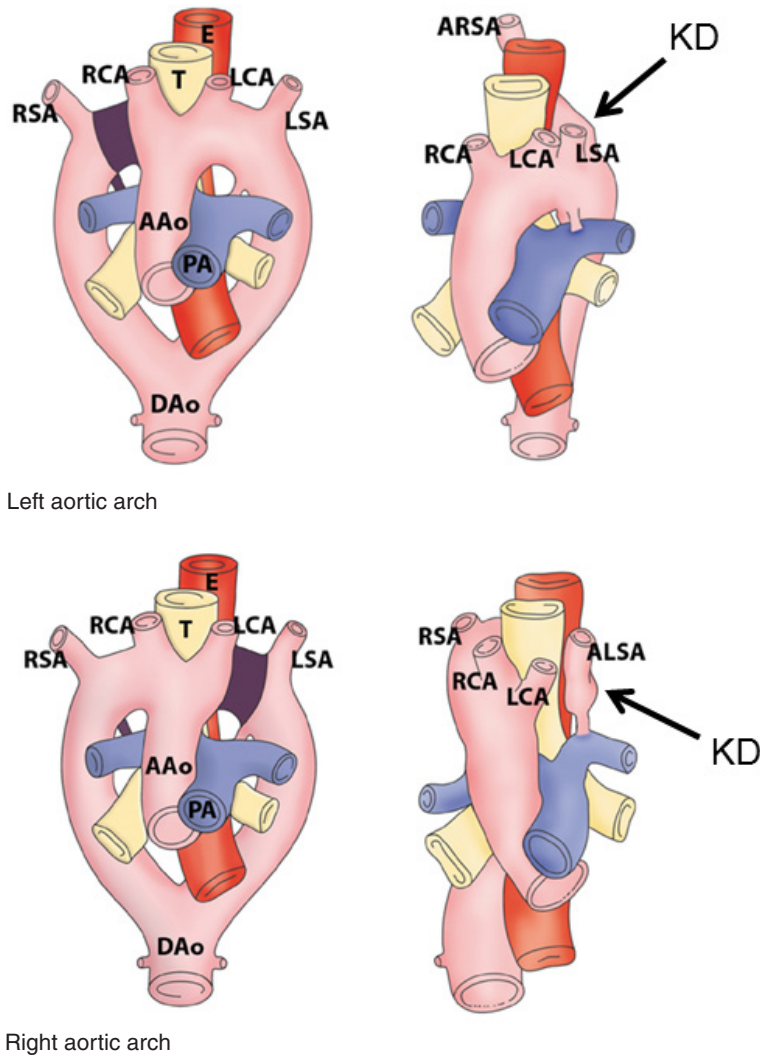


Fig. 4 Schematic drawings of embryology of Kommerell diverticulum: adapted from reference 2.
 RSA: right subclavian artery, RCA: right common carotid artery, LSA: left subclavian artery, LCA: left common carotid artery, AAo: ascending aorta, DAo: descending aorta, T: trachea, E: esophagus, PA: pulmonary artery, ARSA: aberrant right subclavian artery, ALSA: aberrant left subclavian artery, KD: Kommerell diverticulum

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