[CASE REPORT]

A Rare Case of Submassive Pulmonary Embolism with a Right Aberrant Subclavian Artery and Thrombosed Kommerell Diverticulum

Natsuki Onishi, Tomo Komaki, Masayuki Nakamura, Tadaaki Arimura, Joji Morii,
Masahiro Ogawa and Shin-ichiro Miura

Abstract:

An 81-year-old man presented with shortness of breath and was referred to our hospital with suspected acute pulmonary embolism. Enhanced computed tomography revealed a right aberrant subclavian artery with a thrombosed Kommerell diverticulum (KD), as well as deep vein thrombosis in the left leg and bilateral pulmonary artery thrombosis. Thrombosis in the KD disappeared after one month of anticoagulation treatment with rivaroxaban. Thrombosis of a KD is a rare condition that may cause distal emboli and subclavian steal syndrome, although this syndrome was not present in this case. Rivaroxaban is an effective anticoagulant for treating thrombosis of a KD.

Key words: a thrombosed Kommerell diverticulum, pulmonary embolism, a strong hypercoagulable state, rivaroxaban

(Intern Med 59: 1861-1865, 2020)

(DOI: 10.2169/internalmedicine.4495-20)

Introduction

The right subclavian artery normally arises from the innominate artery and crosses anterior to the esophagus and trachea. If present, a right aberrant subclavian artery (ASCA) branches from the aorta as a fourth branch and crosses posterior to the esophagus and trachea. A right ASCA is caused by a developmental abnormality of the aortic arch during embryogenesis and is reported to be present in approximately 1% of individuals (1). If the ductus arteriosus is on the right and a right ASCA is present, the two vessels form a vascular ring around the trachea and esophagus and may compress them. When the origin of the ASCA is dilated to a diameter similar to that of the descending aorta, the dilated site is referred to as a Kommerell diverticulum (KD).

KD was first identified by Burckhard F. Kommerell in 1936 as the cause of dysphagia lusoria (2). It is a rare condition that can be accompanied by medial degeneration of the aorta and carries a risk of aortic dissection and rup-

ture (3-5). Erben et al. reported that a KD grows at a mean (standard deviation) rate of 1.45 (0.39) mm/year and that hypertension is a predictor of growth (6).

In the case presented here, enhanced computed tomography performed to investigate suspected pulmonary embolism (PE) revealed a right ASCA and thrombosis in the KD, which we refer to as a “thrombosed KD.” To our knowledge, only two reports exist regarding a thrombosed KD. We report on the clinical importance and treatment of this condition.

Case Report

An 81-year-old man presented at a nearby hospital with shortness of breath. He had a history of hypertension, dyslipidemia, and hyperuricemia but no history of embolism. He was diagnosed with bronchial asthma, and treatment was initiated with oral medication and an inhaler. However, his symptoms worsened, and at a subsequent consultation he had hypoxemia, with an oxygen saturation (SpO₂) of 92% in room air. A 12-lead electrocardiogram

Figure 1. Echocardiogram. A: the apical four-chamber view, B: the parasternal short axis view. Echocardiogram demonstrates the right-sided dilatation and the D-shaped left ventricle which indicates the right ventricular pressure overload. RA: right atrium, LA: left atrium, RV: right ventricle, LV: left ventricle

Figure 2. Chest X-ray. The X-ray showed no abnormality in the lung field, as well as a left-sided aortic arch.

showed T-wave inversion in the right precordial leads from V1 to V3. Furthermore, an echocardiogram revealed the right-sided dilatation and right ventricular pressure overload with a transtricuspid pressure gradient (TRPG) of 60 mmHg, although the left ventricular ejection fraction was preserved (Fig. 1). Three weeks after his initial consultation, the patient was referred to our hospital with suspected acute PE.

On admission, the patient's heart rate was 94 beats per minute, and his blood pressure was 131/74 mmHg. His hemodynamics were stable. His heart and lung sounds were normal, and no leg swelling was detected. Laboratory tests revealed renal dysfunction, with elevated levels of creatinine (1.21 mg/dL; normal range: 0.65-1.07 mg/dL), D-dimer (4.6 µg/mL; normal range: ≤1.0 µg/mL), and troponin I (36.2 pg/mL; normal range: <30.0 pg/mL). Chest X-ray showed no abnormalities in the lung field and a left-sided aortic arch (Fig. 2). Enhanced computed tomography (CT) detected distal bilateral pulmonary artery thrombosis and left deep vein thrombosis (Fig. 3). We diagnosed him with acute PE and classified it as submassive because of the presence of right

ventricular strain without hemodynamic collapse. In addition to the above findings, enhanced CT detected a right ASCA with a KD at its aortic origin (dilation: 30 mm). The KD was not compressing the trachea and esophagus, but it contained a thrombus (Fig. 4). Although the right subclavian artery was stenosed by the thrombus, the patient had no related symptoms. Carotid duplex ultrasound revealed an antegrade flow in the right vertebral artery, and head magnetic resonance imaging did not show cerebral embolism. We initiated anticoagulant treatment with rivaroxaban 30 mg/day and performed CT 7 days later, which showed a reduction in the thrombi in the pulmonary arteries and KD.

We tested for coagulation factor abnormalities that could cause thrombophilia. His homocysteine level was elevated, but all other values, including antithrombin III, protein S, protein C, and lupus anticoagulant, were in the normal range (Table 1). We performed whole-body CT, checked for tumor markers, and examined the gastrointestinal tract, and all tests were negative for malignancy. The patient's symptoms improved, and he was discharged after 15 days in the hospital. The dose of rivaroxaban was decreased to 15 mg/day after 3 weeks, and the patient continued to receive rivaroxaban in the outpatient department.

At the follow-up evaluation 1 month after discharge, the D-dimer value was negative, and an echocardiogram showed the improvement of the right-sided dilatation with a TRPG of 43 mmHg. Enhanced CT showed no visible thrombosis in the KD, although the thrombi in the distal pulmonary arteries remained (Fig. 5). We suggested operating on the KD to prevent rupture, but the patient did not want to undergo invasive treatment. Therefore, we continued to strictly control his blood pressure and perform regular CT in our outpatient department. However, we explained that an operation may have to be considered if the diameter of the KD increased.

Figure 3. Enhanced computed tomography (CT). Enhanced CT detected bilateral pulmonary artery thrombosis. White arrow shows the thrombus of superior trunk and yellow arrows show the thrombi of lobar arteries.

Figure 4. Volume-rendered image of the aortic arch with supra-aortic vessels. A: anterior view, B: posterior view. C: axial image, D: coronal image. White arrows point to the thrombus in the Kommerell diverticulum. KD: Kommerell diverticulum (black arrow), LSCA: left subclavian artery, RSCA: right subclavian artery, LCCA: left common carotid artery, RCCA: right common carotid artery

Discussion

A right ASCA occurs as a result of abnormal development of the aortic arch during embryogenesis, and dilatation at its aortic origin is called a KD. A KD can compress the trachea and esophagus, leading to respiratory and gastroin-

testinal symptoms, and can also cause aortic dissection and rupture (3). In the case presented here, the KD was not compressing the trachea and esophagus, but it did contain a thrombus.

To our knowledge, a thrombosed KD is rare, and only two other cases have been reported (7, 8). We compared the characteristics of these cases and ours in Table 2 and emphasize the important points as follows: First, the PE was complicated by a thrombosed KD in two of the three cases, and the hypercoagulable state was caused by a paraneoplastic phenomenon in two of the three cases. In the case presented here, we did not detect any malignancy, and the main cause of hypercoagulability was unclear, although the homocysteine level was mildly elevated. Based on the findings of the three cases reported thus far, we suggest that the presence of a thrombosed KD indicates a strong hypercoagulable state and recommend that physicians check for malignancy when they encounter such a state in clinical practice. Second, it is necessary to administer anticoagulation treatment for a thrombosed KD because it carries a risk of distal emboli and subclavian steal syndrome due to vascular occlusion. In the present case, the thrombus in the KD had completely disappeared one month after initiation of rivaroxaban, indicating that rivaroxaban is an effective anticoagulant for treating a thrombosed KD. The other two reports on

Table 1. Coagulation Factors.

	Measurement in patient	Reference range
D-dimer	4.6 µg/mL	0-1
PT	11.8 sec	10.5-13.5
APTT	35.5 sec	24-38
ATIII	>130%	80-130
Protein C activity	100%	70-150
Free protein S level	109%	60-150
LA	1.15	<1.30
Anti-CLβ2GP1 antibody	<1.2	0-3.5
Homocysteine	16.3 nmol/mL	3.7-13.5

PT: prothrombin time, APTT: activated partial thromboplastin time, ATIII: antithrombin III, LA: lupus anticoagulant, Anti-CLβ2GP1 antibody: anti cardiolipin-beta2 glycoprotein 1 complex antibody

Figure 5. Follow-up computed tomography (CT) 1 week and 1 month after initiation of anticoagulant treatment. The thrombus in the Kommerell diverticulum was smaller 1 week after initiation of anticoagulant treatment and had disappeared after 1 month.

Table 2. The Characteristics of Three Cases of a Thrombosed Kommerell Diverticulum.

Reference	Patient characteristics		Subclavian steal phenomenon	Pathogenesis of the hypercoagulable state	Pulmonary embolism	Treatment
	Age	Sex				
(7)	87	M	(+)	Advanced prostate cancer	(+)	Anticoagulant (details unknown)
(8)	70	F	(+)	Non-small cell lung carcinoma	(-)	Anticoagulant (details unknown)
Our case	81	M	(-)	Malignancy (-) Mild hyperhomocysteinemia (+)	(+)	Rivaroxaban

a thrombosed KD stated that anticoagulation treatment was started, but they did not describe the type of anticoagulant used or its effects (7, 8). To our knowledge, this is the first report on the effect of anticoagulant treatment for a thrombosed KD. Because of the risk of aortic dissection and rupture associated with KD, surgical resection may be advisable (3-5). If patients do not agree to surgical resection, strict treatment of hypertension and regular CT examinations are indicated. Finally, this case developed submassive PE, but the thrombi of pulmonary arteries were confined to the distal portion, and the D-dimer level was not remarkably elevated, findings that were inconsistent with the degree of PE. This reminds us of the possibility that the chronic thromboembolic pulmonary hypertension (CTEPH) caused by chronic repetition of thromboembolic event already exists at the baseline, and new thromboembolization at pre-admission exacerbates this condition. Although pulmonary hypertension persisted at one month after discharge, we treated him with an anticoagulant only because he did not complain of dyspnea. If his symptoms recur, we will consider an additional evaluation including ventilation-perfusion lung scanning, right heart catheterization, and pulmonary angiography for the diagnosis of CTEPH.

The authors state that they have no Conflict of Interest (COI).

References

1. Richardson JV, Doty DB, Rossi NP, Ehrenhaft JL. Operation for aortic arch anomalies. *Ann Thorac Surg* **31**: 426-432, 1981.
2. van Son JA, Konstantinov IE, Burckhard F. Kommerell and Kommerell's diverticulum. *Tex Heart Inst J* **29**: 109-112, 2002.
3. Kim KM, Cambria RP, Isselbacher EM, et al. Contemporary surgical approaches and outcomes in adults with Kommerell diverticulum. *Ann Thorac Surg* **98**: 1347-1354, 2014.
4. Doi T, Sakata K, Gyoten T, et al. Two-stage hybrid repair in a patient with acute type A aortic dissection associated with right aortic arch with aberrant left subclavian artery originating from a Kommerell diverticulum. *Ann Vasc Dis* **9**: 223-227, 2016.
5. Wong RH, Chow SC, Lok JK, et al. Hybrid treatment for ruptured diverticulum of Kommerell: a minimally invasive option. *Ann Thorac Surg* **95**: e95-e96, 2013.
6. Erben Y, Brownstein AJ, Velasquez CA, et al. Natural history and management of Kommerell's diverticulum in a single tertiary referral center. *J Vasc Surg. Forthcoming*.
7. Vlummens P, Houthoofd B, Janssens W. Thrombosis of an arteria lusoria with secondary subclavian steal syndrome and swallowing difficulties. *J Belg Soc Radiol* **99**: 80-84, 2015.
8. Faggioni L, Gabelloni M, Napoli V, Iorio F, Chella A, Caramella D. Thrombosis of Kommerell's diverticulum with subclavian steal phenomenon in a patient with non-small cell lung carcinoma under chemotherapy. *Eur J Radiol Open* **3**: 191-194, 2016.

The Internal Medicine is an Open Access journal distributed under the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view the details of this license, please visit (<https://creativecommons.org/licenses/by-nc-nd/4.0/>).