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Case Report

Coronary artery aneurysms of unknown origin in a 14-year-old girl

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ABSTRACT

The case of a 14-year-old girl with acute myocardial infarction due to coronary artery aneurysms (CAAs) of unknown origin, which resembled coronary artery lesions caused by Kawasaki disease, is reported. She was transferred to our hospital due to chest pain with ST-T elevation. She had no history of Kawasaki disease. On the first admission, she was misdiagnosed with acute myocarditis. Then, 54 days later, she experienced chest pain with exertional dyspnea. Her electrocardiogram showed negative *T* waves in the chest leads. A CAA of the left coronary artery was suspected on two-dimensional echocardiography. Coronary angiograms showed 90% stenosis and multiple CAAs of the left anterior descending artery and the bifurcation of the left coronary artery. Both the right coronary artery and left circumflex artery were occluded. A left ventriculogram showed dyskinesis and an aneurysm at the apex. She underwent triple-vessel coronary artery bypass grafting, and her symptoms improved. In addition, an intracranial aneurysm was also found on cerebral angiography. There were no specific laboratory findings other than SS-A antibodies. It was suspected that the weakness of the vessels was related to the disease. It may have been a different disease that was never previously detected, but her CAAs were Kawasaki-like CAAs.

<**Learning objective:** A timely precise diagnosis of acute myocardial infarction is unlikely to be made in children because they are rare. The present patient was initially misdiagnosed as having acute myocarditis, because the coronary arteries could not be detected by two-dimensional echocardiography. Either computed tomographic angiography or magnetic resonance angiography is recommended in patients with ST-T abnormalities on the electrocardiogram if it is difficult to identify the coronary arteries. In this case, the patient had a rare coronary artery disease in which the cause of the coronary artery aneurysms was unknown.

Introduction

The most common cause of coronary artery aneurysms (CAAs) in children worldwide has become Kawasaki disease (KD) [1]. CAAs other than those caused by KD are very rare. It is also known that in those patients with CAAs, in whom it was unknown whether they had a history of acute KD, that the CAAs are presumed to be due to KD. We called these patients presumed KD patients [2]. We describe a case of a young female patient with acute myocardial infarction (AMI) due to CAAs of unknown origin.

Case report

A 14-year-old girl was transferred to our hospital from a nearby hospital because of chest pain with ST-T changes on her electrocardiogram (ECG) and elevation of troponin I level during a long holiday. She had a history of migraine and allergy to pollen. She had no history of vasculitis including KD. Her school-based ECG screening at 12 years of age was normal. On admission, her heart rate was 90 beats per minute, and her blood pressure was 130/80 mmHg. Her heart sounds were regular, and she had no murmur. There were no other abnormal findings on her physical examination. Her ECG showed abnormal *Q* waves in leads II, III, and aV_F and ST-T elevation in V_2-V_4 (Fig. 1 left). The serum creatine kinase (CK), lactate dehydrogenase (LDH), aspartate aminotransferase (AST), and troponin T levels were 1,025 IU/L, 482 IU/L, 127 UI/L,

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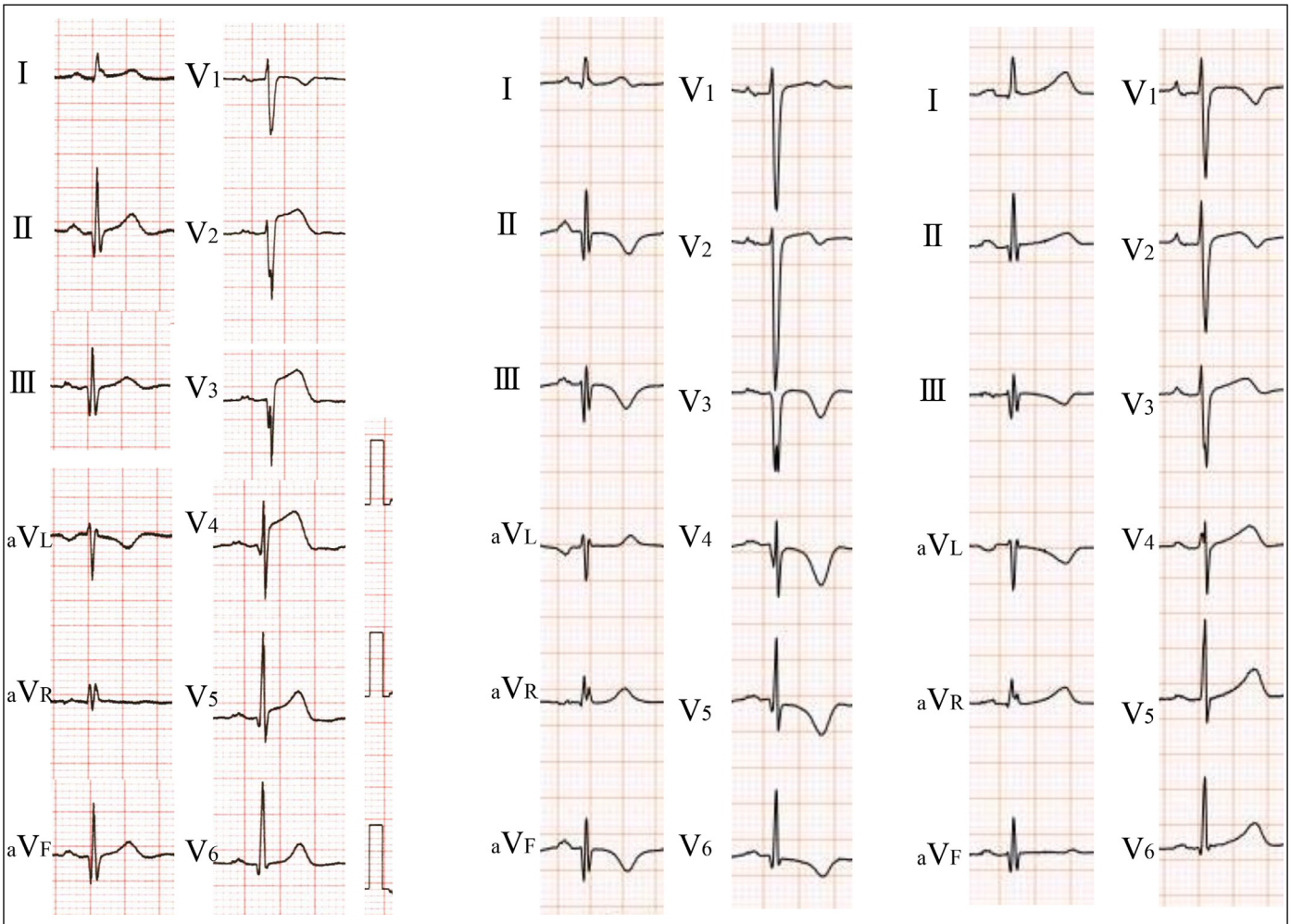

Fig. 1. The 12-lead electrocardiograms at the initial admission, second admission, and postoperative condition. (Left) The initial admission: abnormal *Q* waves in leads II, III, and aV_F and ST-T elevation in V_2-V_4 are observed. (Middle) The second admission: negative *T* waves in leads II, III, aV_F, V_3, V_4, V_5, and V_6 are observed. (Right) 5 months after surgery: negative *T* waves in leads II, V_3, V_4, V_5, and V_6 have improved.

and 0.97 ng/mL, respectively. The white blood cell (WBC) count and C-reactive protein (CRP) concentration were 7.23 × $10^3/\mu$L and 0.35 mg/dL, respectively. The brain natriuretic peptide (BNP) concentration was 74.4 pg/mL. On two-dimensional echocardiography (2DE), the left ventricular ejection fraction (LVEF) was 61%, and the endocardium of the anteroseptal wall was highly echoic. It was difficult to identify the bilateral coronary arteries on 2DE at that time. She was diagnosed as having acute myocarditis. Although she had no fever, she had a fever the next day. The chest pain resolved spontaneously after a few days, and she was discharged on the 10th day after admission. There were no ST-T changes on treadmill testing one month after discharge. Then, 54 days later, she was again admitted due to chest pain, and she also had exertional dyspnea. Her heart rate was 86 beats per minutes, and her blood pressure was 140/84 mmHg. The cardiothoracic ratio on her chest X-ray was 45%. The ECG showed negative *T* waves in leads II, III, aV_F, V_3, V_4, V_5, and V_6 (Fig. 1 middle). The serum CK, LDH, AST, and troponin T levels were 264 IU/L, 209 IU/L, 38 UI/L, and 0.34 ng/mL, respectively. The WBC count and CRP concentration were 6.40 × $10^3/\mu$L and 0.74 mg/dL, respectively. The BNP concentration was 232 pg/mL. The levels of serum antistreptolysin, C3, C4, and CH50 were 227 IU/mL, 147.0 mg/dL, 31.4 mg/dL, and 49.6 U/mL, respectively. The levels of anti-nuclear antibody, anti SS-A antibody, and anti SS-B antibody, were 40 U/mL, 40.2 U/mL, and 2.3 U/mL, respectively. Both rheumatoid arthritis factor and antineutrophil cytoplasmic antibody were negative. Only the anti SS-A antibodies were positive. There were no abnormal findings on the laboratory examination for any specific infections such as Epstein-Barr virus. On 2DE, a CAA of the left main coronary trunk (LMT) was suspected (Fig. 2E).

Cardiac catheterization was performed. The coronary angiogram showed 90% stenosis and multiple CAAs of the left anterior descending artery (LAD) and the LMT (Fig. 2 C, D). The diameter of the CAA in the LMT was 7.2 mm, and that in the LAD was 4.0 mm (Fig. 2E). The right coronary artery was occluded at the orifice, and there were some collateral arteries from the LAD to the posterior descending artery (Fig. 2B, D). The left circumflex artery was also occluded (Fig. 2C, D). Stenosis was also found in the peripheral portion of the high lateral artery. The left ventriculogram revealed dyskinesia and an aneurysm at the apex. The left ventricular end-diastolic pressure was 10 mmHg, and the LVEF was 45%. Computed tomography coronary angiography showed no CAA calcification. Contrast-enhanced cardiac magnetic resonance imaging showed late gadolinium enhancement in the subendocardium of the anterior, septal, and inferior walls and apex of the left ventricle. She was diagnosed as having recurrent myocardial infarction due to CAAs. However, the cause of the CAAs was unknown. An intracranial aneurysm of the vertebrobasilar system was also found

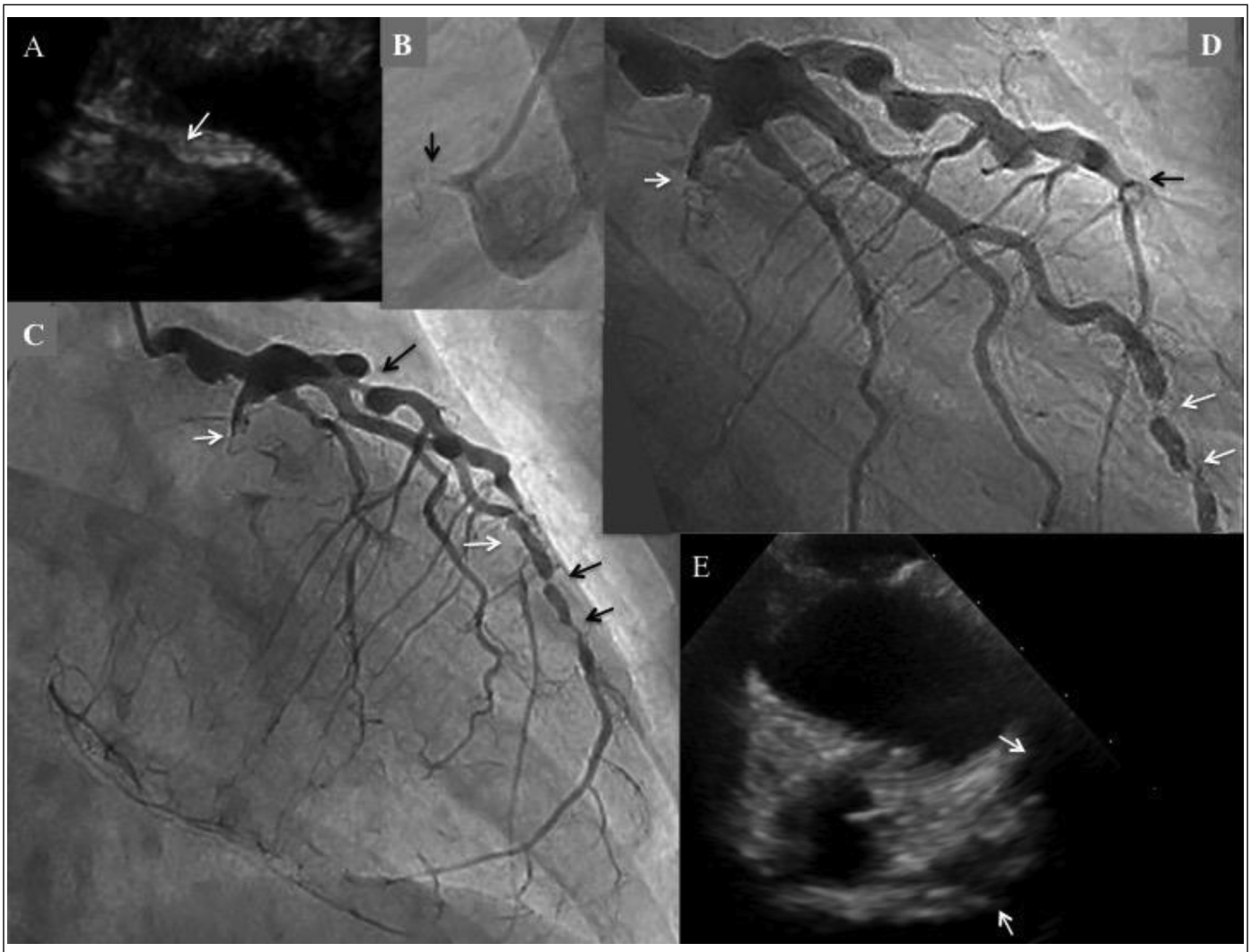

Fig. 2. Coronary angiograms and cardiac echocardiograms. (A) The proximal site of the right coronary artery by two-dimensional echocardiography. (B) Aortic angiogram. (C) Left coronary angiogram. (D) Left coronary angiography. (E) The proximal site of the left coronary artery by two-dimensional echocardiography. The right coronary artery is occluded at the orifice, and there are some collateral arteries from the left anterior ascending artery to the posterior descending artery. The left circumflex artery is also occluded. The left coronary angiogram shows 90% stenosis and multiple coronary artery aneurysms of the left anterior descending artery and left main trunk. Stenosis is also found in the peripheral portion of the high lateral artery.

on brain magnetic resonance angiography, however, there were no aneurysms affecting her trunk or extremities. Cerebral angiography revealed a fusiform aneurysm formed in a rare remnant artery, a persistent primitive lateral vertebrobasilar anastomosis, connecting between the left vertebral artery and middle basilar trunk (Fig. 3). Four months after the onset of the initial AMI, she underwent triple-vessel coronary artery bypass grafting (CABG). She was given warfarin, clopidogrel, carvedilol, and enalapril. After surgery, her exertional dyspnea improved, as well as the negative *T* waves on her ECG (Fig. 1 right). After 1 year, coronary angiograms revealed the patency of the grafts and her coronary arterial lesions were not remarkable changed.

Discussion

The cause of AMI in this patient was thrombotic occlusion due to CAAs. A precise diagnosis of AMI is unlikely to be made in children, because the incidence of AMI is very low in children as compared with adults. In childhood and adolescence, the most common cause of AMI is CAA caused by KD. The detection of coronary aneurysms by 2DE in children is very important, even if the patient has no history of KD. The causes of CAAs are well known, including not only KD, but also incomplete KD [3]. Whether her CAAs were caused of KD or not would be controversial. The patient had no principal symptoms of acute KD. Furthermore, the coronary artery lesions on her CAG were not completely compatible with those caused by KD. CAAs occur primarily in the proximal portions of the major coronary arteries in KD [4]. Her coronary artery lesions appeared to involve the entire coronary arteries, including the peripheral coronary arteries. It seems that her CAAs differed from the coronary artery lesions caused by KD, because of her history and the characteristics of the coronary artery lesions. Therefore, her CAAs would not be diagnosed as being caused by KD.

When did the patient first develop coronary artery disease of an unknown origin? Asymptomatic coronary artery occlusions of the right coronary artery and left circumflex artery may have occurred before the initial episode of the LAD occlusion. There were no abnormal *Q* waves on her school-based ECG screening at 12 years of age. Therefore, it was thought that some coronary events had occurred for 2 years. However, it could not be determined when the CAAs had appeared. Usually, coronary artery calcifications are likely to appear with CAAs after many years, however, that de-

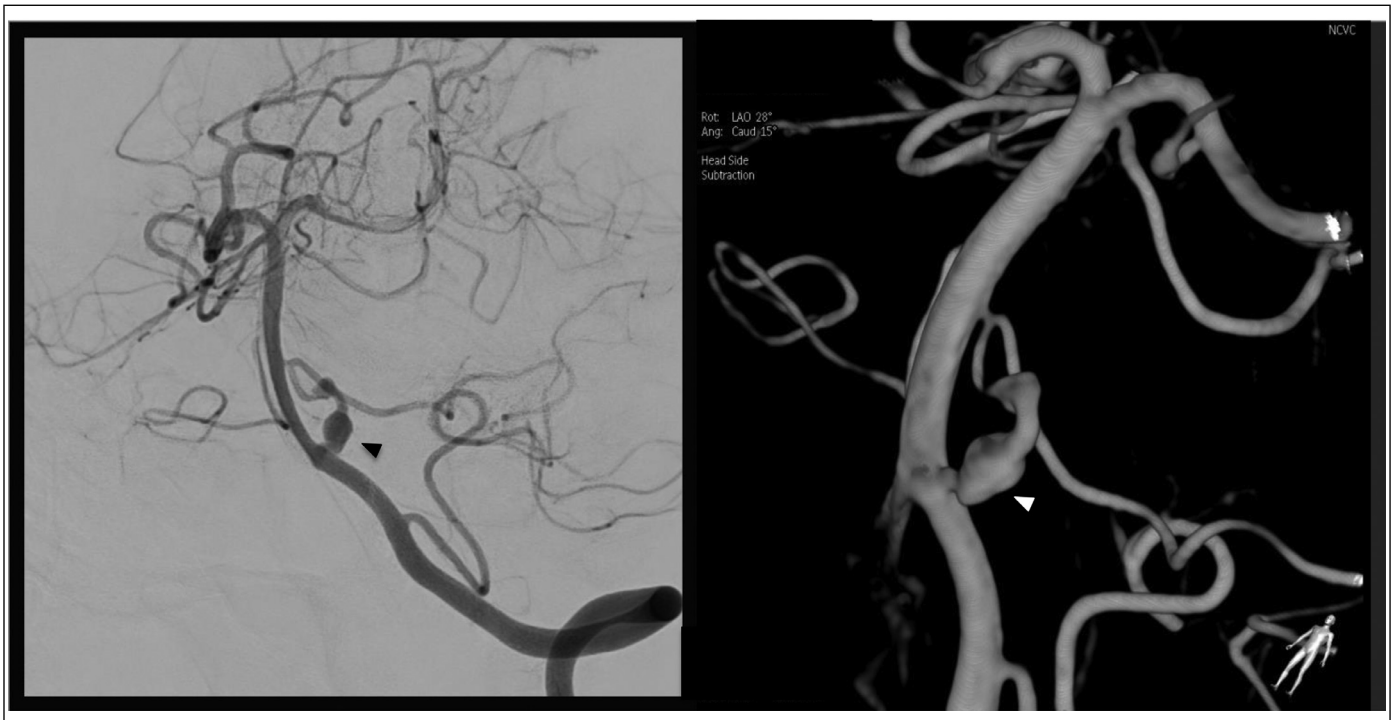

Fig. 3. An intracranial aneurysm associated with a persistent primitive lateral vertebrobasilar anastomosis. (Left) A left vertebral angiography on conventional digital subtraction angiography. (Right) A left vertebral angiogram from 3D digital subtraction angiography. The fusiform aneurysm (arrow head) was formed at a rare remnant artery, a persistent primitive lateral vertebrobasilar anastomosis, connecting between the left vertebral artery and middle basilar trunk.

pends on the characteristics of the underlying disease [5]. One may conclude that she did not have a very long history of CAAs, however, that is just one of several hypotheses.

CAAs are found in 0.3% to 4.9% of patients undergoing coronary angiography [1,6]. The most common etiology of CAAs is atherosclerosis in adults. Furthermore, there are congenital and iatrogenic causes, infections including syphilis, borreliosis, and Epstein-Barr virus infections, connective tissue diseases such as Ehlers Danlos syndrome, Marfan's syndrome, and systemic lupus erythematosus, arteritis such as polyarteritis nodosa, Takayasu's disease, and other causes such as fibromuscular dysplasia. In this patient, it is unknown whether anti SS-A antibodies are related on her CAAs or not. The follow-up in the future should be continued. Recently, several genome-wide association studies have reported genes predisposing to CAAs [7]. At autopsy of patients less than 15 years old in Japan, have been 166 KD cases (77.2%) among 215 systemic vasculitis. There have also been 30 cases (14.0%) with unclassifiable systemic vasculitis cases [8]. It should be recognized that there are some rare cases of CAAs for which the causes are unknown. Whether there was a relationship between the intracranial aneurysm associated with a persistent primitive lateral vertebrobasilar anastomosis and the CAAs in this patient was unknown [9]. However, a weakness of the medium sized arterial wall was suspected. In these CAA diseases, thromboembolisms and ruptures can occur as complications of the CAAs. For the treatment of CAAs, antithrombotic therapies, percutaneous coronary intervention, and CABG are recommended. It has been reported that the prognosis of patients with coronary artery dilatation is poor [10]. Further investigation of the causes of CAAs of unknown origin is needed.

Conclusion

A pediatric case with CAA disease of an unknown origin other than coronary artery lesions caused by KD was presented.

Declaration of Competing Interest

The authors declare that there is no conflict of interest.

Ethical approval

We got consent for this report from her parents.

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