Cervical aortic arch with multiple aortic aneurysms that required two aortic replacements with a 34-year interval

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Abstract

A 57-year old female was referred to our hospital after symptoms of hoarseness and dysphagia for a few years. She was diagnosed with a cervical vessel anomaly during childhood and had undergone surgery for a right-sided thoracic descending aortic aneurysm through a right thoracotomy at the age of 23 years. Now, enhanced computed tomography revealed newly developed multiple aneurysms in the aorta between the left carotid artery and the proximal descending aorta in the left hemithorax. A successful aortic surgical repair was accomplished via a left posterolateral thoracotomy. To our knowledge, this is the first case of a cervical aortic arch with formation of multiple aneurysms that required multiple surgeries via bilateral thoracotomies. This case suggests the inherent fragility and possibility of later aneurysmal formation in malformed vessels, thereby emphasizing the importance of periodical imaging tests in this rare congenital anomaly.

Keywords: Cervical aortic arch • Aneurysm formation • Surgical repair

INTRODUCTION

Cervical aortic arch (CAA) is an extremely rare congenital anomaly. CAA associated with aneurysm formation requiring surgery is an even rarer condition. We report a case of left CAA and the right descending aorta associated with formation of multiple aneurysms that required multiple surgeries via bilateral thoracotomies.

CASE REPORT

A 57-year old female was admitted to our hospital for further evaluation and surgical treatment of aortic aneurysms. She was diagnosed with anomaly of cervical vessels during childhood. She underwent surgery for thoracic aortic aneurysm in her right-sided descending aorta via a right thoracotomy at the age of 23 years. Although she had had no symptoms for years since the first surgery, she started to have hoarseness and dysphagia several years before the current admission.

On physical examination, the patient was found to have a pulsatile left supraclavicular mass. Blood pressure was 130/60 and 100/60 mmHg in her right and left arms, respectively. A chest radiograph revealed a superior mediastinal shadow with calcification, extending to her left neck (Fig. 1A). Enhanced computed tomography (CT) revealed left-sided CAA with the right-sided descending aorta. The branching pattern of the three major arteries was normal. Three saccular aneurysms were present between the left carotid artery and the left sub-clavian artery, and one of the aneurysms involved the orifice of the left sub-clavian artery. Another aneurysm was located in the proximal descending thoracic aorta in the left hemithorax (Fig. 1B). The aorta beyond the fourth aneurysm ran transversely over the vertebral column, and its distal descending thoracic aorta, which included a piece of vascular prosthesis implanted in the previous surgery, was right-sided.

OPERATIVE FINDINGS

Surgery was performed through a posterolateral thoracotomy in the left fourth intercostal space to approach the distal aortic arch and the thoracic descending aorta. She also had a persistent superior vena cava. After systemic heparinization, the left femoral artery and vein were cannulated, and partial cardiopulmonary bypass was established. The aortic arch was then clamped just distal to the left carotid artery and the descending aorta distal to the fourth aneurysm. The left sub-clavian artery was clamped. The aneurysm was incised and the aorta was reconstructed in the anatomical position with a 14 mm prosthetic graft (J Graft Shield Neo; Japan Lifeline, Tokyo, Japan). The left sub-clavian artery was reconstructed to the side of the aortic graft using an interposed 7 mm prosthetic graft. The operation time and cardiopulmonary bypass time were 320 and 81 min, respectively. Histological examination of the resected specimen revealed atherosclerotic change and thinning of the medial layer of the arterial wall. (Fig. 2B). The postoperative course was uneventful, and she was discharged on the 28th postoperative day.
DISCUSSION

CAA is an extremely rare congenital anomaly that comprises cephalic displacement of the aortic arch. Aneurysms develop in \( \approx 20\% \) of cases [1]. To our knowledge, 23 cases of surgically treated CAA with aneurysms have been reported [1–5]. The condition of a left-sided CAA in conjunction with the right descending aorta is even rarer, with only 4 cases reported [1–5]. The unique feature of this case, in contrast to other reports, is that formation of multiple aneurysms required multiple surgeries via bilateral thoracotomies at remotely different times.

Although most CAAs are asymptomatic, when complicated by aneurysmal dilatation, patients may have symptoms of dysphagia, dyspnoea or frequent pulmonary infection, which are caused by compression of the oesophagus and trachea. Compared with common atherosclerotic aortic aneurysms, aneurysms associated with CAA usually occur in relatively younger patients (the youngest was aged 6 years) [1].

Although the embryogenesis of CAA remains unclear, it is speculated that abnormal persistence of the third brachial arch and the regression of the fourth arch, which normally forms the left aortic arch, could be involved [2]. The aetiology of aortic aneurysmal dilatation in CAA remains unclear. Possible explanations for it include abnormal embryologic development of the aortic wall, connective tissue, abnormal haemodynamics and aortic wall stress.

In the present case, aortic replacement was performed twice via a bilateral thoracotomy. In the first operation, the site of the aneurysm was the right descending thoracic aorta. In the second operation, it was the transverse aortic arch and the proximal descending aorta that were replaced with a prosthetic graft; all these sites of the aorta were embryologic remnants normally designated to regress. It is unlikely that the aneurysmal formation was caused by the aortic cross-clamp in the previous surgery, because the site of each operation was remote. Histological examination of the resected specimen revealed atherosclerotic change and thinning of the medial layer of the aortic wall, which suggested a congenital weakness. This inherent fragility and abnormal haemodynamics may have contributed to aneurysm formation. In general, CAA develops an aortic aneurysmal dilatation in the early phase. If a remnant of the malformed vessel remained in place after the operation, this site of the aorta may become dilated later in life.

Figure 1: Preoperative images. (A) Chest X-ray shows the aortic arch deviating cranially over the left clavicle. (B) 3D-CT shows saccular aneurysms between the left carotid artery and the proximal descending thoracic aorta in the left hemithorax. (C) The arrow indicates that one of the aneurysms compresses the oesophagus. (D) The arrow head reveals that the thoracic aorta runs transversely over the vertebra. Asterisk indicates the left superior vena cava.
Therefore, we suggest that postoperative periodical medical check-ups by imaging tests are necessary in these cases.

CONCLUSION

We report a very rare case of CAA in conjunction with the right descending aorta with multiple aneurysm formation that required multiple surgeries via bilateral thoracotomies at remotely different times. The malformed vessels in this disease seem to be inherently fragile; therefore, postoperative follow-up by periodical imaging tests is recommended to detect developing aneurysms.

Conflict of interest: none declared.

REFERENCES