Late Aneurysm of the Distal Aortic Arch after
Repair of Aortic Interruption

A Case Report

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Summary

Aneurysm formation after aortic coarctation repair is not a rare complication of post-coarctation of aorta repair. We describe the case of a 43-year-old woman who had undergone repair of an isolated interruption of the aortic arch 30 years earlier, who came to our hospital with progressive chest pain, cough and dyspnea. A giant aortic aneurysm was revealed in the distal aortic arch by CT study. The patient underwent aneurysmectomy with total aortic arch replacement using a Dacron graft through redo median sternotomy. An embryologic explanation of this patient’s anomaly and the previous surgical procedure are discussed for defining this rare clinical condition. (Jpn Heart J 1999; 40: 497–501)

Key words: Isolated interruption of the aortic arch, Embryologic pathogenesis, Late aneurysm formation, Total aortic arch replacement

PSEUDOANEURYSM formation in the long term is by no means rare in cases with a history of aortic coarctation (Co/Ao) repair which used graft or patch materials. Direct end-to-end anastomosis has recently become the choice of primary surgical treatment for Co/Ao or interruption of the aortic arch (IAA). In this article, we report the case of a giant aneurysm of the distal aortic arch in a patient who had a history of repair of an isolated IAA using an artificial graft 30 years earlier.

Case Report

A 43-year-old woman, who 30 years earlier had undergone interruption repair by graft interposition with a 12 mm woven Teflon tube graft at another
institution, was admitted to our hospital after 3 years of progressive dyspnea which had worsened with coughing and chest pain over the last month before admission. A grade 2/6 systolic ejection murmur was heard loudest on the upper left sternal border. The right upper-extremity blood pressure was 110/66 mmHg and the lower-extremity blood pressure was 90/70 mmHg. A chest X-ray was obtained, showing aortic arch enlargement (Figure 1). Computed tomography

**Figure 1.** Anteroposterior chest X-ray. Presence of an aneurysm of the aortic arch shown in a standard X-ray film. The left diaphragm was elevated due to phrenic nerve palsy.

**Figure 2.** 3D-CT of the chest. A: This huge aneurysm was noted on the distal aortic arch by viewing from the caudal side. B: The arch vessels were clearly visualized by viewing from the cranial side. The left subclavian artery originated from the top of the aneurysm (arrow). AS = ascending aorta; G = old graft; AN = aneurysm; D = descending aorta.
(CT) of the chest revealed a 6.5 cm mass adjacent to the distal aortic arch. This huge aneurysm originated in the distal aortic arch around the old graft anastomosis and the left subclavian artery originated from the aneurysm (Figure 2). The IAA between the left common carotid artery and the left subclavian artery was not associated with intracardiac defects or with a patent ductus arteriosus (PDA). This type of IAA has been reported only rarely and had not been classified according to the Celoria-Patton classification of IAA. The patient had no history of anti-hypertensive medication after IAA repair.

Emergent aneurysmectomy was performed due to the progressive chest pain. The surgical approach to the aneurysm was through a redo median sternotomy for total aortic arch repair. Deep hypothermic circulatory arrest with cerebral perfusion was applied. When the aorta and former prosthetic graft were opened, no mural thrombus was noted in the dilated proximal descending aorta. No aneurysmal dilatation of the woven Teflon tube graft was noted. No fibrous strand connecting the ascending aorta, the descending aorta and the pulmonary artery was found during dissection around the aortic arch. Aortic continuity was successfully reestablished by a total replacement of the aortic arch using a collagen coated 16-mm Dacron graft (Hemashield: Meadox Medicals, Inc., NJ, USA). The postoperative course was uneventful. The coughing and chest pain were relieved by aneurysmectomy. The patient was discharged 3 weeks after operation and is doing quite well 2 years postoperatively.

DISCUSSION

Infants with the combined lesions of IAA, PDA and ventricular septal defect can survive only days or weeks without surgical intervention. On the other hand, isolated IAA not accompanied by a PDA or intracardiac anomaly is known as a rare malformation with a favorable clinical course. Most of these patients can live through childhood and into adolescence due to an effective collateral supply to the descending aorta. There are reports that elective operative procedures have been successfully undertaken using direct anastomosis or interposition of an artificial graft. Although direct anastomosis should be the first choice of surgical repair for IAA, the interposition of a prosthetic graft may be needed to bridge the interrupted aortic arch. The present case had been reported as one of the first successful cases of surgical repair with a graft interposition in 1967. It is also well known that late pseudoaneurysm formation at the coarctation repair site is a rare, but serious complication, if patch aortoplasty has been used for the Co/Ao repair. There was no evidence of anastomotic aneurysms with obvious dehiscence of textile-aorta anastomoses in this case.

Late aortic aneurysms related to a congenital anomaly of the great vessels
Figure 3. The possible embryologic pathogenesis of this anomaly. Abnormal regression of the left fourth aortic arch (IV: shadowed) and normal regression of the left side dorsal aorta (RDA) were considered to be the embryologic genesis in this patient. The left sixth aortic arch (VI) was regressed and the ductus arteriosus was not patent. Ao = aorta; PA = pulmonary artery; IA = innominate artery; RCA = right common carotid artery; LCA = left carotid artery; RDA = regressed dorsal aorta; LSA = left subclavian artery.

should be treated after consideration of the genesis of these lesions. The possible embryologic explanation of this patient’s defect is pictured in Figure 3. There was an abnormal regression of the left fourth aortic arch and a normal regression of the left-sided dorsal aorta between the left third and fourth arches, which explain the interruption of the aortic arch between the left common carotid artery and the left subclavian artery. It is possible to say that this anomaly was categorized into Celoria-Patton type B IAA with normal regression of the left sided dorsal aorta. From the point of view of embryology, her aortic arch was thought to be constructed without persistent embryologic components, which should normally be regressed in the course of embryo generation.

As another possible cause of this aneurysm, hypertension after surgical treatment for Co/Ao and a post-stenotic (small diameter prosthetic graft in childhood) aneurysm could also be considered. However, the precise pathogenesis could not be defined.

Three dimensional CT (3-D CT) has been widely used and is noted as a valuable tool in evaluating aneurysms of the thoracic aorta and in confirming the relation between aneurysms and cervical branches. It is also useful for classifying IAA types and for obtaining information about previously implanted prosthetic grafts. In this case, precise information about the location of the aneurysm and the origination of the arch vessels was clearly visualized by 3-D CT prior to redo
surgery (Figure 2). Rupture of pseudoaneurysms after Co/Ao repair have sometimes been reported to be fatal. Thus, once a diagnosis has been established by CT study, repair should be performed as soon as possible.

**Conclusion:** An aneurysm around the reconstructed aorta should be considered in the differential diagnosis for patients with chest pain and a history of repair of congenital aortic anomaly. A rare case of late aneurysm formation around the site of previous repair of an isolated IAA was presented. Long term follow-up including CT study is necessary for these patients. Aggressive surgical treatment is definitely indicated as soon as the diagnosis of an aneurysm of the thoracic aorta is established.

**References**