Left Cervical Aortic Arch With Aortic Coarctation and Saccular Aneurysm

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Cervical aortic arch is a very rare malformation and is occasionally accompanied by other cardiovascular anomalies. A 48-year-old male patient had a left cervical aortic arch with aortic coarctation and saccular aneurysm distal to the coarcted segment. The major clinical manifestations were upper body hypertension with a 50-mmHg discrepancy between the upper and lower limbs and a loud continuous murmur in the upper chest and back. Magnetic resonance angiography successfully depicted the anomalous aorta, and the aortic coarctation and aneurysm were surgically resected and the thoracic aorta was reconstructed. The discrepancy in blood pressure diminished after the operation, but antihypertensive medication was continued to satisfactorily control the hypertension. (Jpn Circ J 2000; 64: 544 – 546)

Key Words: Aortic coarctation; Aortic saccular aneurysm; Cervical aortic arch

Aortic arch anomalies are reported to occur in 0.5 - 3% of patients! Among them, cervical aortic arch (CAA) is a developmental entity in which the aortic arch is cranial to its usual position! with right CAA occurring slightly more often than left CAA. Less than 60 cases of CAA have been reported in the English literature to our knowledge and, in most cases, it is an isolated condition and usually clinically silent. However, CAA may be associated with other cardiovascular anomalies! Clinical features vary from a pulsatile mass in the supraclavicular fossa to dysphagia or chronic respiratory diseases.

The conventional method for the diagnosis of CAA is transarterial angiography, but noninvasive means such as magnetic resonance angiography (MRA) are more desirable! We report the first case of left CAA accompanied by aortic coarctation and saccular aneurysm just distal to the coarcted segment.

Case Report

A 48-year-old male patient was referred to The Institute for Adult Diseases Asahi Life Foundation Hospital for examination and treatment of hypertension in June 1997. In his childhood history there had been an abnormal shadow observed in his upper mediastinum and an abnormal murmur in his left upper chest. Since then the patient had developed normally, but had been kept under observation. At age 30, hypertension was diagnosed at an annual health check-up, but the patient declined further evaluation and treatment. He had no history of significant diseases, such as aortitis or infective endocarditis, and no symptoms of chronic respiratory infection or swallowing disorder.

In June 1997 the patient presented with an intractable headache and was referred for further evaluation. On physical examination, his blood pressure was 172 / 100 mmHg in both upper limbs and 100 / 62 mmHg in both lower limbs. A loud, continuous murmur was present in the left upper chest and back and a thrill was palpable under the left scapula. Routine blood examination was normal: The standard serologic test for syphilis was negative and the white cell count, C-reactive protein, and erythrocyte sedimentation rate were all within the normal range.

A chest X-ray showed that the aortic arch was deviated...
Fig. 2. Aortic coarctation located just distal to the ostium of the left subclavian artery and a saccular aneurysm just distal to the coarcted segment. (A) Magnetic resonance angiography. AAo, ascending aorta; DAo, descending aorta; A, coarcted segment; B, saccular aneurysm. (B) Transarterial digital subtraction angiography. BC, brachiocephalic artery; LCC, left common carotid artery; LSC, left subclavian artery; A, coarcted segment; B, saccular aneurysm.

Fig. 3. (A) Surgical findings: arrow A indicates the coarcted segment; arrow B indicates the thin-walled saccular aneurysm. (B) Resected specimen: arrow A indicates severe stenosis with marked calcification in the coarcted segment; arrow B indicates the very-thin-walled saccular aneurysm.
cranially over the clavicle with a normal cardiac silhouette and without rib notching (Fig 1). An electrocardiogram showed normal sinus rhythm and left ventricular hypertrophy with ST depression. An echocardiogram revealed a normokinetic left ventricle with moderate concentric hypertrophy and no valvular or congenital abnormality. Finally, gadolinium-enhanced MRA was performed, which revealed a left CAA with ipsilateral descending aorta and the major 3 arteries branching normally (Fig 2A). It also revealed that an aortic coarctation was present just distal to the left subclavian artery, with a saccular aneurysm 3 cm in size located distal to the coarctation and a tortuous descending aorta beneath the aneurysm. Consequently, the patient was transferred to the University of Tokyo Hospital for further evaluation and possible surgical treatment.

Transarterial catheterization was performed, which showed normal coronary arteries and an anomalous aorta similar to the MRA findings (Fig 2B). A peak-to-peak pressure across the coarctation was approximately 50 mmHg. As it was feared that aneurysm may rupture, and for treatment of the upper body hypertension, the coarcted segment and saccular aneurysm were resected and the descending thoracic aorta was reconstructed with a woven Dacron graft. The ductal ligament, however, was not identified during surgery. The postoperative course was uneventful and the discrepancy in the blood pressure between the upper and lower limbs diminished. However, antihypertensive medication was continued to maintain normal blood pressure.

Histopathological examination of the resected specimen (Fig 3) showed severe stenosis of the aorta with marked intimal hyperplasia and calcification. The saccular aneurysm had a very thin wall with secondary defects of the intima and media, indicating a false aneurysm at high risk of rupture. There was no sign of leukocyte infiltration or granulomatous change around the narrowed segment or the aneurysm.

Discussion

This is the first reported case of CAA complicated by aortic coarctation and a saccular aneurysm, although there has been previous reports of CAA accompanied by aortic coarctation or a fusiform aneurysm. CAA is a very rare congenital aortic anomaly of unknown origin, possibly caused by the persistence or confluence of the higher branchial aortic arch or by the failure of caudal migration of a normally formed aortic arch. Aortic coarctation is generally thought to be caused by constriction of the ductal ligament, because the aorta at the attachment of the ductus arteriosus comprises ductal tissues, a hypothesis supported by an angiography study in Japanese. However, the cause of the aortic coaractation accompanying CAA might be different because it has been reported in unusual situations: one case had diffuse aortic narrowing located in an unusual position and another had multiple coarctations.

Aortic coarctation is sometimes accompanied by fusiform or saccular aneurysms which may be congenital or secondary due to the hemodynamic disturbance caused by the coarctation. A previous report described the occurrence of coarctation-related aneurysms on the lower-pressure side as almost as frequent as those on the higher-pressure side which suggests a congenital weakness of the arterial wall. Inflammation of the aorta may be also the cause of these aneurysms, such as Takayasu arthritis or infective endocarditis. In the present case, there was no evidence of inflammatory change or infection in the surgical specimen, so it is more likely that the saccular aneurysm formed secondarily to the turbulent blood flow passing through the coarctation.

Magnetic resonance imaging or angiography (MRA or MRA) can reliably define many different aortic arch anomalies without invasive procedures or exposure to radiation. The distinct advantages of MRI and MRA over other imaging modalities, such as computed tomography and transarterial angiography, are the ability to image in different planes, the comparatively large field of view, and excellent spatial and contrast resolution. We performed both transarterial catheterization and MRA for the evaluation of the anomalous aorta and believe that MRA has the advantage because of its noninvasive nature.

In summary, we report the first case of CAA complicated by aortic coarctation and a saccular aneurysm. MRA successfully depicted this anomalous aorta, indicating the usefulness of this noninvasive method in the diagnosis of aortic anomalies.

References


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