Anomalous Right Subclavian Artery Associated with Postductal Aortic Coarctation: Case Report

POSTDUKTAL AORT KOARKTASYONUNDA GÖRÜLEN ANORMAL SAĞ SUBKLAVİAN ARTER

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Summary

Here we present the case of a young man with anomalous right subclavian artery and coarctation of the aorta. If anomalous right subclavian artery originates from the distal portion of the aortic coarctation it will be major collateral for the descending thoracic aorta. In our case, there was a cerebral flow steal by the vertebral artery to the upper extremity and/or descending thoracic aorta. Distal aortic pressure was sufficient (50-55 mmHg) while the anomalous right subclavian and left subclavian arteries were clamped. So that no additional procedure to preserve spinal cord was performed. Patient had no complication postoperatively and he is at his first follow-up year, without any clinical problems.

Key Words: Anomalous right subclavian artery, Aortic coarctation, Major collateral, Cerebral flow steal

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Özet

Sunduğumuz erkek hastada anormal sağ subklavyan arter ve aort koarktasyonunun birlikteliği söz konusu idi. Eğer anormal sağ subklavyan arter aort koarktasyonunun distal kesiminden orijin almaktaysa descendan torasik aorta için major kollateral damardır. Bizim vakamızda vertebral arter yoluyla üst ekstremite ve/veya descendan torasik aortaya olan serebral akımda steal bulunmaktaydı. Anormal sağ subklavyan arter ve sol subklavyan arter klemplendiğinde distal aortik basınç yeterli bulunmuş (50-55 mmHg) ve spinal kordu korumaya yönelik ilave bir prosedür uygulanmamıştır. Hastanın postoperatif komplikasyonu olmadı ve takibinin 1. yılında olup herhangi bir klinik problemi bulunmamaktadır.

Anahtar Kelimeler: Anormal sağ subklavian arter, Aort koarktasyonu, Majör kollateral, Serebral akımda steal

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The anomalous right subclavian artery (ARSA) is the most common abnormality of the left aortic arch development with a frequency of 0.5 %(1). First described by Hunauld(2) and an incidence of 1 % of an ARSA associated with a postductal aortic coarctation was reported by Maude Abbott (1).

Several types of congenital vascular anomalies produce partial or total encirclement of the trachea or esophagus frequently leading to dysphagia or dyspnea with stridor or both. (2) Aneurysms in the course of an ARSA are not frequently encountered and may develop such clinic in some infants or young children from the retroesophageal course of aberrant artery. Description and treatment by ligation of ARSA was made by Gross in 1946 (3).

Here we present the case of a young man with ARSA and coarctation of the aorta in which ARSA was the major collateral for the descending thoracic aorta.

Case Report

A 20 years old man admitted to our outpatient clinic with complaints of rhinorrhage and palpitation. He had deafness and dumbness due to meningitis in childhood. On physical examination there was a systolic murmur in left sternal area. There were palpable and pulsatile vessels in parascapular and subscapular area and a mild systolic murmur could be heard. Peripheral pulses were palpated weaker at lower extremities. Arterial pulses were also weaker in right arm when compared to the left (Table 1).

Table 1. The comparison of arterial blood pressures preoperatively.

	Preoperative	**Postoperative**
Upper extremities	Left 160/70, Right 90/60 mmHg	Left 120/60, Right !25/65 mmHg
Lower extremities	Left 90/55, Right 85/50 mmHg	Left 120/65, Right 120/60 mmHg

Figure 1. Digital substraction angiography (DSA) and aberrant right subclavian artery arising from the distal portion of the coarctated segment.

Figure 2. Magnetic resonance imaging (MRI) demonstrating the postductal aortic coarctation.

In the chest X-ray, the heart was enlarged and notchings were detected only at the left side on the thirth and fourth rib. The transthoracic echocardiography (TTE) revealed a segment of coarctation in descending aorta distal to left subclavian artery and the measured peak gradient was 70 mmHg in the coarctation area. Digital substraction angiography (DSA) supported the diagnosis.Here arrows show aberrant right subclavian artery arising from the distal portion of the coarctated segment (Figure 1). These findings were visualised also with magnetic resonance imaging (MRI) and catheterisation (Figure 2). Esophagography showed no compression (Figure 3).

Operation

After monitorization of both radial arterial blood pressures, left posterolateral thoracotomy through the fourth intercostal space was performed and coarctation was exposed. Distal aortic arch, isthmus, descending aorta and ARSA arising from the arch was dissected (Figure 4). The vessel distal to the coarctated segment was estimated as the ARSA. There was no pressure wave for the right

Figure 3. Esophagography showed no compression.

Figure 4. Aortic arch, isthmus, left subclavian artery and ARSA was dissected.

Figure 5. The estimated vessel as ARSA was proved by a guide wire introduced from right radial artery which was seen in the orifice of estimated vessel distal to coarctation.

radial artery first, however, when the ARSA was clamped the blood flow was directed through the vertebral artery to the right arm. And distal aortic pressure was 60/30 mmHg. The distal aortic pressure when ARSA and left subclavian artery were clamped was 50-55/30mmHg measured with an arterial line and intracath. This intracath was kept in place to measure distal aortic blood pressure during cross clamping. With this pressure measurements, it was decided that the circulation of spinal cord was sufficient and no additional protective procedure was needed. When the clamp was released, the blood flow was directed to the descending thoracic aorta. The ligamentum arteriosum was ligated. Coarctated segment included proximal left subclavian artery partially, so left subclavian artery was not suitable for clamping. We then clamped arcus aorta proximal to the origin of left subclavian artery, ARSA, left subclavian artery and descending aorta distal to coarctated segment. Longitudinal aortotomy was made through the distal side of left subclavian artery up to normal aorta, along the coarctated segment. The estimated vessel as ARSA was proved by a guide wire introduced from right radial artery which was seen in the orifice of estimated vessel distal to coarctation (Figure 5). Well-developed ridge of coarctation was partially resected. Aortic patchplasty with 6 x 4 cm ovale shaped Dacron graft was performed (Figure 6). Even though the ARSA was a major collateral for descending thoracic aorta, the distal aortic pressure was thought to be sufficient while clamping ARSA and left subclavian artery and no additional procedure to preserve spinal cord was performed during operation. After declamping there was no pressure difference between right and left radial arteries and distal aorta.

Figure 6. Aortic patchplasty with (a sealed Dacron graft Meadox) ovale shaped graft was performed.

No complication occured perioperatively. Postoperative blood pressures were measured as 120/70 mmHg in both upper extremities and 125/75 mmHg in lower extremities. Postoperative DSA demonstrated the surgical correction of the coarctation was succesful and ARSA was running in the previous course (Figure 7). Patient was discharged on the postoperative 6th day without any complication.

Figure 7. Postoperative DSA demonstrated the surgical correction of the coarctation was succesful and ARSA was running in the previous course.

Discussion

The normal right subclavian artery has three embryological components; the most proximal segment is derived from the fourth arch; the middle segment is produced by the cranial portion of the right dorsal aorta and the most distal segment is developed from the right seventh intersegmental artery. During normal embryological development right dorsal aorta disappears between the origin of the seventh intersegmental artery and the junction with the left dorsal aorta (1).

An ARSA arising from the descending thoracic aorta is thought to result from persistence of the right dorsal aorta and disapperance of the right fourth arch and cranial portion of the right dorsal aorta. Thus, the ARSA as it originates from the descending thoracic aorta and pass across the mediastinum posterior to the esophagus, is derived from the right dorsal aorta and right intersegmental artery. Knowledge of the embryological development of this anomaly would suggest that its point of origin is on descending thoracic aorta distal to ligamentum arteriosum (a remnant of the left sixth aortic arch) and therefore distal to the site of the usual location for postductal aortic coarctation (1,4,5). When, however coarctation is associated with an ARSA, the anomalous vessel may be found proximal or distal to the coarctation site or ARSA may rise from the coarctation site (1,4).

ARSA is only common in type B aortic arch interruption (AAI), but is rare in type A and coarctation of the aorta. Absolutely, ARSA is the rule in type B AAI. Such high incidence of ARSA might be due to the hemodynamic alterations. Reduced flow through the right fourth arch, is responsible, since the arch is also supplied by the ascending aorta (6). In our case, there was approximately 3-4 cm of segmentary aortic coarctation with a raphe in the mid region and the ARSA originated distal to the raphe.

In the intraoperative evaluation, the abnormal vessel was proven to be the ARSA with a guide wire introduced from the radial artery inthrough the vessel to the estimated orifice. Meanwhile, prior to clamping of ARSA, there was no arterial wave in the right radial artery seen on monitor. However when the ARSA was occluded near the coarctation segment, the arterial pressure waves of right radial artery appeared and after releasing the clamp, there was no sign on the monitor again. This was probably because of the collateral circulation. When the ARSA was clamped, the blood flow from vertebral artery was sufficient only for the right subclavian therefore the arterial pressure wave was seen. However, when the clamp was released, the blood flow was directed to the descending thoracic aorta, as it is less resistant, the arterial pressure waves disappeared. Even though the ARSA was a major collateral for descending thoracic aorta, we clamped and repaired the coarctation with a 6x4 cm hemashield patch graft.

In childhood, ARSA is pliable and flexible. However, in adults, there exists rigidity, tortiosity and dilatation according to atherosclerosis. Compression over vena cava superior, trachea or esophageous may be seen (2). In our case, we did not see such complications due to localisation and size. As the ARSA was originated from the distal side of the raphe, the pressure was low and there was no tortiosity or aneurysmatic dilatation, there was only an enlargement in the calibration. Other potentially lethal complications such as aneurysmatic dilatation, rupture and/or distal embolus are the

main indications for surgery in terms of diagnosis (3). The higher frequency of serious complications of anurysms such as hemorrage, distal embolisation or rupture dictates that ARSA should be operated promptly upon diagnosis. Clearly available data indicate that excision should be done soon after the diagnosis is made unless other diseases indicate a serious operative risk (3).

The ARSA can be used as a flap (subclavian flap method) to correct coarctation of the aorta. Easy to perform, potential of growth and low morbidity, mortality rates are the advantages for the suitable patients (1). When ARSA is used as a flap, it will also treat the obstructive or compressive symptoms to the esophagus or trachea. Using ARSA as a flap deals with both problems.

In patients with an aortic coarctation and normal development of the arch vessels and in those with an ARSA proximal to coarctation, the internal mammarian artery, subscapular branches provide important collateral pathways for blood flow beyond coarctation. When an ARSA originates distal to the coarctation, this vessel acts as the main collateral channel. Retrograde blood flow occurs from the right vertebral artery into the subclavian arteries and descending thoracic aorta (1). In our case, there was no clinical symptom for cerebral blood flow steal, as flow of the contralateral side was sufficient.

ARSA originated from the distal portion of the aortic coarctation, is a major collateral for the descending thoracic aorta. In our case, there was a cerebral flow steal by the vertebral artery to the upper extremity and/or descending thoracic aorta. In spite of the intraoperative examination, no additional procedure or preservation was performed as the distal aortic pressure was sufficient while the ARSA was clamped. The patient is at his first follow-up year, without any clinical problem.

Most techniques for repair of coarctation of the aorta require temporary occlusion of the thoracic aorta. The proximal effect of increased vascular resistance on the heart and particularly the distal effects of ischemia of the spinal cord are minimised during occlusion by the presence of collateral vessels. In patients with aortic coarctation and ARSA undergoing surgical correction, importance and cognisance should be taken of the role of the anomalous artery in providing collateral flow to the descending aorta. If the ARSA arises distal to the coarctation, it may act as the main collateral channel for descending aorta and clamping should be avoided (1). In these patients there is an increased risk for postoperative complications such as paraplegia due to spinal cord ischemia. Techniques such as atriofemoral bypass, ventriculofemoral bypass, jump grafts or systemic hypothermia are advised to prevent complications. If the anomalous vessel is distal to the coarctation, care should be taken to prevent spinal cord ischemia (1). In our case, no paraplegia did occur although ARSA was distal to the coarctated segment. The factors effecting this situation is the age of our patient, well-developed collateral circulation, palpable femoral artery pulses and distal aortic pressure was sufficient to protect spinal cord from ischemia while ARSA and left subclavian artery is clamped.

Although there was no problem postoperatively in our patient, besides ARSA, left subclavian artery clamping can increase the risk of spinal cord injury. Therefore, distal aortic blood pressure and sufficiency of collateral circulation should be investigated thoroughly preoperatively.

If left subclavian artery is to be clamped, surgeon should be aware of the risk of paraplegia and follow distal aortic blood pressure cautiously. If distal aortic blood pressure is thought to be not sufficient, other appropriate protective methods of spinal cord shoul be used to prevent ischemia.

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