Anomalous Innominate and Right Subclavian Arteries Associated with Coarctation of Aorta

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Anomalies of the aortic arch and its branches comprise a wide variety of conditions giving rise to varied symptoms and signs. Most of these anomalies are amenable to surgical correction and relief of symptoms. This paper reports a case with four distinct vascular anomalies: anomalous innominate and right subclavian arteries and coarctation of the aorta with the anomalous subclavian artery emerging from the aorta at the site of the coarctation.

History and Incidence

According to Stauffer and Pote, Bayfield, in 1789, reported the first case of a right subclavian artery originating as the fourth branch of the aortic arch. He reported in detail the clinical and postmortem findings. In 1934 a total of 260 cases had been reported in the literature. More recently, Gross and associates began the surgical attack on this condition and reported 11 cases, 10 of which were of aberrant right subclavian arteries and one of an aberrant left subclavian artery. All patients survived operation and were relieved of their symptoms.

Stauffer and Pote report the incidence of the right subclavian artery originating as the last branch of the aorta as being anywhere from 0.4% to 2% of anatomic dissections, and our review corroborates these findings. Anomalies of the right subclavian artery are found most frequently in association with some other vascular

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malformation. Brean and Neuhauser, in 1947, saw three cases of anomalous right subclavian artery associated with patent ductus arteriosus. Kroeker et al. reported a coarctation with anomalous right subclavian artery and subaortic stenosis. McGregor and Medalie have seen an 8-year-old patient with coarctation and anomalous right and left subclavian arteries. In 1915 Love and Holms performed a necropsy on a patient who had coarctation with a stenotic right subclavian artery coming off the innominate artery, the innominate and left common carotid arteries originating from a common trunk. Dolgopol's studies reveal the independent origin of the right subclavian artery to be the third commonest anomaly of the arch, the first being an innominate artery which gives off a right subclavian and the two carotid arteries, with the left subclavian coming off separately. The second commonest anomaly is that in which four branches are seen to emerge from the aorta, the fourth branch being the left vertebral artery. An independent right subclavian may be the first, second, or third branch but most frequently is the fourth.

The incidence of coarctation of the aorta is reported at 0.1%. We were unable to find any reference in the literature as to the incidence of an innominate artery arising from a left aortic arch, although a left innominate artery arising from a right aortic arch is not uncommon. Likewise, we were unable to find a reference in the literature substantiating a case of aberrant right subclavian artery coming off the aorta at the level of the coarctation, al-
though cases have been reported in which the anomalous subclavian arises just above or below the coarctation.⁵, ²⁴ Thus, the combination of these conditions—coarctation of the aorta, aberrant right subclavian artery arising at the site of coarctation, and left innominate artery—must be exceedingly rare, and we can find no reference to such a case in the English literature.

**Embryology**

An anomalous right subclavian artery is caused by an abnormal obliteration of the right fourth aortic arch, forcing the right subclavian to come off the aorta distal to the left subclavian. From a developmental point of view, the first part of an anomalous right subclavian artery is not the right subclavian at all.¹¹ Rather, it is the dorsal portion of the right aortic arch, and Renander²³ believes that cases of this anomaly are correctly classified as instances of right aortic arch.

The embryology of coarctation of the aorta is very poorly understood, and the reader is referred to an excellent review of this subject by Edwards et al.¹² A left innominate artery with a normal aortic

Fig. 1.—Posteroanterior roentgenogram of the chest, showing the hazy mass in the left upper mediastinum.
arch is embryologically possible, but we have found no reference to it in the literature; it would be caused by a failure of obliteration of the left ventral aorta between the third and fourth aortic arches, along with an obliteration of its fellow on the opposite side.

**Report of a Case**

The patient, a 28-year-old white man, was admitted to the hospital for treatment of a supposed peptic ulcer. A lifelong history of leg claudication on exercise was present. There was nothing else significant in the past history.

Physical examination revealed a man of small stature. The blood pressure was 160/85 in the left arm and 103/70 in the right arm. In both lower extremities the blood pressure was approximately 100 systolic. The chest was clear to percussion and auscultation. Large pulsating vessels were palpable on the left posterior chest wall. A Grade 3 systolic murmur was heard over the entire precordium but was best heard in the pulmonic area, this murmur being transmitted to the neck and to the posterior chest to the left of the midline. The right radial pulse was weaker than the left. The femoral pulses were reduced in volume and compared roughly with the right radial pulse. The pulses in the lower extremities were not obtainable. No other significant abnormalities were
found, and no symptoms referable to esophageal compression were present. The laboratory revealed a normal hemogram and urinalysis. Chest x-ray showed a density in the left upper mediastinum (Fig. 1), and the trachea was pushed forward, suggesting a mass behind it. There was moderate scoliosis in the midthoracic region of the spine with the convexity to the right. A retrograde aortogram showed a coarctation and the left innominate artery with no evidence of subclavian filling on the right (Fig. 2). Tortuous internal mammary and thoracic vessels were present. The EKG was normal. Fluoroscopy indicated that the heart was somewhat enlarged, and a pulsating mass was seen in the left portion of the upper mediastinum, producing a deviation of the esophagus to the right. Operation was performed through a left posterolateral incision. The coarctation was seen at the isthmus, with a somewhat atretic, anomalous right subclavian artery coming off at the site of the coarctation on the posterior aspect of the aorta, this vessel running superiorly and posteriorly behind the esophagus (Fig. 3). A ligamentum arteriosum came off the medial aspect of the aorta at the level of the coarctation. The vessels in the chest wall, the intercostal arteries, and the internal mammary arteries were greatly dilated and tortuous. The left subclavian and left common carotid arteries were seen to come off an innominate vessel which had its origin just proximal to the coarctation. The anomalous right subclavian artery was divided at the site of the coarctation; the coarcted segment was excised, and an end-to-end anastomosis was performed, with no tension on the suture line. Postoperatively, the patient did very well, the pulses in the lower extremities becoming bounding to the extent that the dorsalis pedis pulsations were visible. There was no change in the character of the right or left radial pulse. The blood pressure in the left arm became 110/75, in the right arm 90/65, and in the lower extremities 128/80. Three weeks after operation the patient was sent home on convalescent sick leave. After being home for three weeks, he developed hemoptysis and returned to the hospital. A chest film taken at this time revealed a mass, the size of a tennis ball, at the site of aortic anastomosis. A second operation,
using a partial left-heart by-pass, was carried out, and the anterior suture line in the anastomosis was found to be disrupted, thereby creating a false aneurysm. This area was excised and a homograft sutured in place to bridge the gap. Convalescence was uneventful, and the patient was discharged from the hospital six weeks later.

Comment

An aberrant right subclavian artery arises from the posteromedi al surface of the first part of the descending aorta and courses posterior to the esophagus in nearly all cases, although in some instances it may pass between the esophagus and the trachea, or even anterior to the trachea. The treatment of this condition, "dysphagia lusoria," with the complete relief of symptoms, is predicated upon the division of such an aberrant vessel, which compresses the esophagus and/or the trachea. These facts have been well documented by Gross et al., In none of the surgically treated cases, including ours, in which the anomalous right subclavian artery was transected, has circulatory embarrassment to the right arm resulted. The arteries of the right side of the thorax furnish a collateral circulation from the aorta to the axillary artery by way of the subscapular branch, as well as other vessels. Pattison reported seven cases of anomalous right subclavian artery, four of which were associated with a tetralogy of Fallot. Recognition of this condition is important, since Bahnson and Blalock reported that on three occasions they inadvertently used a carotid artery, rather than a subclavian, to perform a shunt operation.

Multiple anomalies of the aortic arch and its branches are not uncommon, but the case presented in this paper showed a combination of four anomalies, which we are unable to duplicate from a careful search of the English literature.

The occurrence of a leak at the anastomotic site following operation is difficult to explain. The aorta was very clean and showed no atheromatous degeneration in this region, and the anastomosis was accomplished under absolutely no tension, with a simple running over-and-over suture of 0000 vascular silk. There were no leaks upon release of the clamps. Leaks at the suture line have been reported, but they commonly occur within two weeks following operation. The leak in our case occurred after six weeks.

Summary

A case with four separate vascular anomalies of the aortic arch is presented.

Recognition of such anomalies is important when one is contemplating any type of surgical procedure on the great vessels. Anomalies of the aortic arch are commonly multiple. If one anomaly is found, others should be looked for.

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REFERENCES


