Double Aortic Arch Associated with Coarctation of the Aorta: Surgically Treated Patient

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Anomalies of the aortic arch system may assume many different anatomic patterns. Some are only of academic interest; others are of extreme clinical importance and may at times constitute an emergency requiring prompt surgical intervention to save life.

The anomalies of the aortic arch system which produce symptoms and which require surgical treatment are of three types. First, there are the "vascular rings," which are so disposed as to form a ring around or to press against the trachea, the esophagus or both; second, those anomalies which produce obstruction in one or another part of the aorta, namely coarctation; and third, persistent patent ductus arteriosus.

The surgical correction of coarctation of the aorta has become a standard procedure and is ideally performed on young adults; occasionally this anomaly produces cardiac symptoms in early life necessitating surgical treatment to avert progressive, at times rapidly progressive, heart failure.

The patient on whom data are reported here had the unusual combination of a vascular ring in the form of a double aortic arch with stenosis of the right (posterior) arch and coarctation of the left (anterior) arch.

Report of Case

A man, 25 years old, came to the Mayo Clinic in November 1951, for surgical treatment of coarctation of the aorta, a condition diagnosed elsewhere 10 years previously. Hypertension had first been discovered during the course of a routine examination in 1937 when he was 11 years old. He had enjoyed excellent health and even though he knew about the hypertension and its cause, he was active in sports which included four years of college football. The patient was unusually well developed, being 6 feet (about 183 cm.) in height and weighing 205 pounds (about 93 kg.). The blood pressure in the right arm was 204 mm. of mercury systolic and 100 diastolic, and in the left arm 210 systolic and 104 diastolic. No pulsations could be felt in the abdominal aorta or in the femoral arteries. There was a systolic murmur over the base of the heart without any appreciable increase in the size of the heart clinically. A routine roentgenogram of the thorax revealed questionable "notching" of ribs (Figure 1). Roentgenoscopic examination revealed some enlargement of the left ventricle. During this examination it was noted that the esophagus was displaced anteriorly, apparently by a retro-esophageal vessel. The electrocardiogram showed a rate of 72, sinus rhythm, notched QRS waves in lead III, notched P waves in lead II, biphasic P waves in lead III, inverted T waves

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in leads III and V-1, positive T waves in leads V-2, V-3 and V-4 and positive T waves and small Q waves in leads V-5 and V-6.

It was appreciated then that, while we were dealing with a coarctation of the aorta, there were present also other anatomic deviations from the normal, not usually seen in cases of coarctation of the aorta. Routine laboratory examinations contributed no additional significant information.

Although it was appreciated that this was not a perfectly typical instance of coarctation of the aorta we felt that exploration of the lesion with resection of the stricutured segment of aorta and anastomosis, if feasible, should be carried out. Accordingly operation was performed on November 24, 1951, by one of us (Clagett). A long curved incision was made around the tip of the scapula and carried high enough posteriorly to permit resection of a long segment of the fifth rib and a short segment of the fourth rib. The patient was a large, remarkably well-developed athlete with powerful shoulder girdle muscles. As usual in cases of coarctation there was an extensive collateral circulation throughout the site of incision.

When the pleura was opened, the findings were typical for coarctation of the aorta, with the stricuture of the aorta involving a left aortic arch and occurring about 2 cm. below the origin of the left subclavian artery and about 0.5 cm. below the ligamentum arteriosum. The internal mammary arteries and intercostal arteries were large and tortuous. On first inspection this was a perfectly typical coarctation of the aorta. We proceeded with dissection. The first three left intercostal arteries below the stricuture were ligated and divided, and the ligamentum arteriosum was ligated and divided. As dissection was carried around behind the aorta it became apparent that there was a large vessel arising from the posterior wall of the aorta just below the site of coarctation. It was first assumed that this was a large right intercostal artery. Further dissection disclosed, however, that this vessel extended upward and posteriorly and that it was, in fact, the posterior or right arch of a double aortic arch (Figure 2). Occlusion of this vessel did not affect pulsation of the right radial artery. It was a large vessel, almost as large as the anterior aortic arch; however, it narrowed at its junction with the descending thoracic aorta. The presence of this vessel seriously complicated any attempt to resect the coarctation and perform an end-to-end anastomosis. At first we

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**FIGURE 1a**

*Figure 1a: Roentgenogram of thorax.*

**FIGURE 1b**

*Figure 1b: Esophagogram.*
considered leaving this vessel undisturbed and proceeding with resection of the coarctation and anastomosis. However, in the course of dissection and mobilization the retro-esophageal vessel ruptured at its junction with the aorta. The proximal end was therefore ligated, and the aortic wall was repaired. The coarctation was then resected, and an end-to-end anastomosis of the aorta was performed.

The segment of aorta removed at operation had an appearance similar to that in the usual case of aortic coarctation in which resection is done for this disease.

FIGURE 2a: Operative area. L.A. = left aortic arch. R.A. = right aortic arch. D.A. = descending aorta. The first three left aortic intercostal arteries have been ligated and divided. The coarctation, involving the left arch, is partly hidden by the stump of the first aortic intercostal artery.

FIGURE 2b: Diagrammatic interpretation of findings at operation.
The specimen measured 1.2 cm. in length. Two millimeters distal to the aortic insertion of the ligamentum arteriosum there was a concavity involving the superior, anterior and posterior walls of the aorta. The wall into which the ligamentum inserted was not deformed. The external diameter of the aorta at the level of the deformity was 0.9 cm. Corresponding to this deformity seen externally was a diaphragm-like membrane across the aortic lumen. Grossly no lumen could be identified at this point.

Microscopic examination revealed the typical deformity of aortic coarctation (Figure 3). The media of the superior, anterior and posterior portions of the aortic wall protruded as a curtain into the lumen of the vessel and constituted part of the diaphragm observed grossly. The basic cause of the luminal narrowing was the deformity of the aortic media. Superimposed on this layer was laminated connective tissue containing elastic tissue. This tissue, which is considered to be acquired tissue, added in a minor degree to the narrowing of the lumen caused by the medial change. No lumen could be identified in serial sections of the specimen.

The patient's postoperative course was uneventful, and he was dismissed from the hospital on his twelfth postoperative day and from our care two days later. He was advised to restrict his physical activities for three months and permanently to avoid strenuous physical exertion. Whereas his brachial blood pressure had been recorded as more than 200 mm. of mercury systolic and more than 100 diastolic preoperatively, his postoperative pressures while in the hospital were recorded as 160 mm. of mercury systolic and 90 diastolic. His abdominal and femoral pulsations were readily felt postoperatively.

FIGURE 3: Longitudinal section of aortic segment removed during operation. Picture is characteristic of coarctation. The lower wall of the aorta, which receives the ligamentum arteriosum (ligamentum art.), is regular in contour. The superior wall shows characteristic infolding of the media, (M), producing the basic narrowing of the lumen. Considerable superimposed intimal tissue (I) is proximal to the coarctation and at the coarctation. This tissue, which in part contains laminated layers of elastic tissue, narrows the lumen in excess of that caused by the medial deformity and obliterates the lumen (Verhoeff's elastic tissue stain and counterstained with van Gieson's connective tissue stain, x 3½).
Comment

Persistent patency of the ductus arteriosus is not an uncommon companion of coarctation of the aorta, occurring in about 10 per cent of clinically recognized examples of coarctation. Other significant malformations of the great vessels in association with coarctation, of which the case herein reported is an example, are, however, rare.

This case of double aortic arch with coarctation of the left arch and stenosis of the right arch has some resemblance to two of the necropsied cases on which data have been analyzed by Arkin.1 In each of these the roentgenologic evidence had suggested the presence of a double aortic arch, a condition verified by necropsy. In one of Arkin's cases the left arch had a short atretic segment beyond the left subclavian artery and proximal to the ligamentum arteriosum. The right arch was widely patent, not showing any point of stenosis like that exhibited in our case. The atretic segment of the left arch in Arkin's case may have represented coarctation; but, since no report of the microscopic appearance of this segment of the left arch was given, it is impossible to state that the atretic portion of the left arch had the same developmental significance as in our case. In our case the narrow segment of the left arch had the characteristic microscopic appearance of coarctation.2

In Arkin's other case in which necropsy was performed there was also a narrow segment involving the left arch in the same location as in his first case. Here the narrow segment was almost 1 cm. long, a fact which makes it unlikely that coarctation which would fulfill existing microscopic criteria existed here.

Regardless of the nature of the narrow segment involving the left component of a double aortic arch in each of the two of Arkin's cases mentioned, it is to be emphasized that the right arch was widely patent and therefore the clinical features of coarctation were not present in his cases.

It is pertinent to mention another, and yet rarer, malformation which may occur with coarctation of the aorta and yield signs of esophageal compression. This is the condition in which the right subclavian artery arises anomalously as the fourth branch of the aorta and below the coarctation. The artery then passes behind the esophagus to reach the right side of the body. A case of this type was represented in the review of Fawcett3 (case 12), and a similar case of Stephens was mentioned by Gross.4

More recently, Sealy5 described a similar case (case 1), the patient being a 19-year-old man, in which the anomalous right subclavian artery was demonstrated at operation. The coarctation of the aorta was proximal to the origin of the anomalous artery. The right subclavian artery was divided, and the segment of aorta containing the coarctation was resected followed by an end-to-end anastomosis of the aorta. It is of passing interest that as the right subclavian artery was clamped preparatory to division, and before the aortic coarctation was disturbed, the blood pressure in the right arm rose. As in the type of case herein reported there may be esophageal compression, but in the cases with the anomalous right subclavian
artery there are weak right radial pulses and low right systolic brachial
blood pressures. In contrast, in our case with a double aortic arch the
blood pressure and pulses in the two upper extremities were approximately
equal.

Comment should be made about the absence of tracheal or esophageal
symptoms in our patient with a vascular ring in the form of a double
aortic arch. Many reports have emphasized the fact that this type of
aortic deformity may give rise to alarming symptoms of respiratory ob-
struction in such cases and may cause death unless the ring is interrupted
surgically. At the same time there are reports of patients living a long
and asymptomatic life with the same type of malformation. Whether or
not symptoms of tracheal or esophageal obstruction result from a vascular
ring probably depends on the degree of compression of these two tubes
by the vascular ring. As a rule, if symptoms occur, they appear in infancy
or childhood. If no disturbances occur during this period, the vascular ring
as such is usually of no concern to the patient. This was the case in the
patient on whom data are presented. The reason for operation was evidence
of intrinsic obstruction to the aortic channel rather than obstruction by
the anomalous vessels to the structures they surrounded.

SUMMARY

The case of a 25-year-old man with double aortic arch, coarctation of
the left arch and stenosis of the right arch is reported. The usual clinical
signs of coarctation of the aorta were exhibited. A vascular malformation
in association with the coarctation was suspected from the roentgenoscopic
examination, which revealed esophageal compression in the upper part of
the thorax. No symptoms of esophageal or tracheal dysfunction were present.

Treatment consisted in resection of the area of coarctation in the left
arch with end-to-end anastomosis of this arch. The continuity of the right
arch was interrupted. The postoperative course of the patient was good.

RESUMEN

Se refiere un caso de un hombre de 25 años de edad con doble arco aórtico
y estenosis del arco derech co. Los signos habituales de la coartación de la
aorta estaban presentes. Se sospechó una malformación vascular asociada
da la coartación por el examen radiológico, que reveló compresión del esófago
en la parte superior del tórax. No había síntomas de transtorno funcional
del esófago o de la tráquea.

El tratamiento consistió en resección del área de la coartación del arco
izquierdo con anastomosis término-terminal de este arco. La continuidad
del arco derecho fue interrumpida. La evolución postoperatoria del enfermo
fué buena.

RESUME

Les auteurs rapportent l'observation d'un homme de 25 ans chez lequel
existe un double arc aortique; il s'y assoie une coarctation de l'arc gauche
et un rétrécissement de l'arc droit. On y constatait les signes cliniques
habituels de la coarctation aortique. L'examen radioscopique faisait envisager une malformation vasculaire associée à la coarctation. Il montrait en outre une compression oesophagienne dans la partie supérieure du thorax. Il n'y avait aucune manifestation clinique oesophagienne ou trachéale.

Le traitement consista en la résection de la zone de coarctation dans l'arc gauche, avec anastomose termino-terminale de cet arc. On supprima la continuité de l'arc droit. Les suites opératoires furent bonnes.

REFERENCES