Multiple and Infra ductal Coarctations of the Aorta

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With the recent rapid improvements in methods of diagnosis and treatment of cardiovascular diseases, the surgeon today can attempt to cure the unusual as well as the more common types of defects. Refinements in the technics of cardiac catheterization, angiography, grafting, hypothermia, and extracorporeal circulation have been a great stimulus to the description of the rare lesions in the hope that ultimately a completely corrective procedure can be performed. In this paper, a case of multiple coarctations of the aorta is presented and discussed with a review of the literature. To our knowledge this is the first case report in which there have been 4 distinct coarctations of the aorta.

Case Report

The patient was a 32-year-old Puerto Rican woman with a known murmur since the age of 12 and a recent onset of precordial discomfort, dyspnea, and fatigue. No signs or symptoms of congestive heart failure or subaeute bacterial endocarditis were evident. Physical examination revealed the following positive physical findings: the blood pressure was, in the right arm 120/90; in the left arm 205/115, and in the right leg 140/100; the right radial pulse was weak, the left was strong, and the femoral pulses were decreased, but palpable. The heart was enlarged to the anterior axillary line in the sixth intercostal space. A grade-II systolic murmur was heard over the entire precordium, loudest along the left sternal border. No collateral pulsations were felt.

Laboratory Findings. The electrocardiogram indicated left ventricular hypertrophy. Radiographie and fluoroscopie examination of the chest showed considerable enlargement of the heart to the left, giving an appearance of left ventricular enlargement. The arch of the aorta showed a shallow impression on its superior posterior aspect and the descending aorta was located unusually far toward the left with the esophagus also somewhat deviated to the left. The configuration of the upper portion of the aorta was somewhat suggestive of the so-called atypical coarctation; however, no narrow segment of aortic arch was visualized, nor was there any evidence of rib notching. Because of the x-ray findings, venous angiography was performed, which revealed a narrowing of the descending aorta in the midthoracic region.

Hospital Course. A diagnosis was made of a lower thoracic aortic coarctation with separate occlusion of the right subclavian artery. At surgery, a left thoracotomy revealed a marked stenosis approximately 2½ inches below the origin of the left subclavian artery. No inflammation or adhesions were observed surrounding the aorta; however, peculiar calcification of the wall of the aorta was noted distal to the coarctation. The aorta was cross clamped above and below the area of coarctation, whereupon the blood pressure rose to over 300 mm. Hg. This blood pressure elevation was controlled with a slow intravenous drip of trimethaphan camphorsulfonate (Arfonad) until the resection of the coarctation was completed. Upon slow removal of the cross clamps, the blood pressure fell markedly and an infusion of levaterenol (Levophed) was instituted. Over the next 15 minutes the Levophed was gradually slowed as the blood pressure rose to about 200 systolic. As the chest was being closed, ventricular fibrillation developed and persisted despite all attempts at resuscitation.

Postmortem Examination. The pericardium and epicardium revealed several areas of hemorrhage and some ﬁbrin secondary to cardiac massage. The heart weighed 570 Gm., and was markedly enlarged due to hypertrophy of the left ventricle. The right atrium and ventricle were comparatively small, so that they appeared as appendages of the left heart. The tricuspid, pulmonic, mitral, and aortic valve rings measured 9.5, 5.5, 9.0, and 6.0 cm., respectively, in circumference. The right and left ventricular myocardium measured 0.3 and 2.5 cm., respectively, in thickness. The valvular architecture and myocardium were otherwise unremarkable.

The aorta was markedly sclerotic and calcified from the region of the transverse arch distally (fig. 1). There was an old arteriosclerotic occlusion in the right subclavian artery at the bifurcation of the innominate artery. An intact fresh annular suture line was present in the aorta 13 cm. distal to
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the aortic valve ring at the site of the first coarctation. A small annular cushion producing only minimal narrowing of the lumen of the aorta was noted immediately below the orifice of the celiac artery. A slightly more prominent ridge was found 18 cm. distal to this area immediately above the orifice of the right renal artery. The intervening segment of aorta was slightly dilated. A severe deformity of the aorta with eccentric narrowing of the lumen produced by a diaphragm-like septum was found 4.8 cm. distal to the origin of the right renal artery. The only communication between the proximal and distal aorta in this region was a channel that extended along the posterior wall of the aorta from a point 1.5 cm. proximal to the septum. This channel measured 0.6 cm. in diameter. The segments of aorta distal to the ridge and diaphragm were moderately dilated. The left renal artery was traced to the region between the ridge and diaphragm, where it ended blindly. No collateral renal arteries were noted. There was minimal atherosclerosis distal to the channel. The iliac, right and left common carotid, and left subclavian arteries measured 1.4, 3, 2, and 2 cm., respectively, in circumference. The brachiocephalic arteries were markedly sclerotic. The coronary arteries were widely patent throughout, with little sclerosis. There was a dimple in the aortic wall at the site of the ligamentum arteriosum.

Fig. 1. Photograph (left) and drawing (right) of aorta opened showing marked atherosclerosis, surgical anastomosis (A), 3 areas of coarctation (B, C, and D) channel (E), and right renal artery (F).
The lungs were partially atelectatic and congested. A few fibrous tags found in the pulmonary arteries were thought to be old organized pulmonary emboli. The liver and spleen were congested. Numerous depressed stellate subcapsular scars and fibrous adhesions were noted over the anterior surface of the liver near the dome of the right lobe. The left kidney weighed 55 Gm. and its surface was smooth. The right kidney weighed 100 Gm. and had numerous vascular scars over the surface.

On microscopic examination the myocardium showed focal scarring. The aorta was markedly atherosclerotic with papillary calcific excrences in areas. The right subclavian and left renal arteries had old occlusions. On section, small arteries were noted adjacent to the left renal artery. The surgically excised segment of aorta consisted of several fragments showing severe atherosclerosis. The sites of coarctation revealed subintimal fibrous thickening and fragmentation of the elastica. The media was thickened in the region of the proximal cushion, and resembled an abortive infolding (figs. 2-5). The ridge revealed more marked infolding. The distal coarctation and channel revealed infolding and approximation of the media to the most marked degree.

The liver was congested. Numerous granulomata were seen scattered throughout the hepatic parenchyma. These varied from hyalized nodules to fibrillar nodules some of which contained ova of Schistosoma mansoni. Many similar lesions were found in the submucoosa of the large and small intestine and lung.

Review of the Literature

Gross has stated that 98 per cent of all coarctations are located in the first segment of the descending aorta just distal to the arch. More recent reports testify to an increasing awareness of coarctations below the isthmus.

Coarctations of the thoracic aorta below the isthmus have been reported as early as 1835, when Schlesinger described a localized narrowed area just above the diaphragm. Another case of a lower thoracic aortic constriction was published by Hasler in 1911. In that report the coarctation was located 3 cm. above the diaphragm, and the aorta was replaced by a fibrous cord for a distance of 2 cm. In 1930 Costa discussed a case with an intra-aortic diaphragm in the lower thoracic aorta. Hickl, in 1931, presented an instance in which there was a cylindrical narrowing just below the isthmus. In 1933, Hahn reported a localized aortic narrowing at the level of the diaphragm. Olim in 1949, attempted to resect a coarctation below the isthmus. His patient was a 20-year-old girl with a lesion located 5 cm. above the diaphragm. The operation failed be-
cause of lack of a graft to replace the 1.7 cm. area that was resected. Bahnson and co-workers,8 in 1949, reported a lower thoracic coarctation 4 cm. in length. The only procedure performed was a sympathectomy. In 1950, Freeman and colleagues9 demonstrated by retrograde carotid aortography, a narrowing of the aorta in the regions of the eleventh thoracic and first lumbar vertebrae. Their patient was a 14-year-old girl with multiple congenital anomalies and a blood pressure of 225/125 in the right arm and 130 in the leg. A successful resection of the lower thoracic coarctation was accomplished by Beattie and associates,10 in 1951. He resected a 6.5 cm. coarctation above the diaphragm and replaced it with a homograft. Brock and Graham,11 in 1952, resected a coarctation of the lower thoracic aorta that was associated with an aneurysm above the area of stenosis. Both lesions were removed and replaced with a homograft. Another successful operation was performed by Glenn et al.,12 in 1952. In their case, the coarctation involved the lower third of the thoracic aorta and the upper abdominal aorta up to the celiac axis. A splenectomy was done and the splenic artery anastomosed to the thoracic aorta above the coarctation. In 1953, Deterling13 resected a long coarctation of the inferior thoracic aorta and replaced it with a graft with good result. Since then, cases have been described by Depraz,14 Dubost and Binet,15 Hulting,16 and Maurea.17 Coarctation of the abdominal aorta is also being seen and reported more frequently. Quain,18 reported the first case of this type in 1847. His case had a narrowing of the aorta just below the renal arteries. In 1861, Power,19 described the lesion immediately below the inferior mesenteric artery. Nothing further was published until the 1930's, when papers by Maycock20 and Baylin21 appeared describing a stenosis below the renal arteries. In 1941, Steel22 reported a 46-year-old woman who died of a cerebral vascular accident and at postmortem examination, a marked constriction of the aorta just above, and at the level of the renal arteries, was found. In 1949, Bahnson et al.,8 published a case in which a preoperative diagnosis of coarctation of the abdominal aorta just below the renal arteries was made by angiography. At operation the aorta was dilated below the diaphragm for several inches and then became quite narrow at the level of the second lumbar vertebra. The adventitia was stripped from the renal artery. The omentum was placed against the left kidney and a bilateral sympathectomy was done. Kondo and associates23 reported a 12-year-old girl who died in acute pulmonary edema. The abdominal aorta, at autopsy, showed a marked degree of stenosis between the celiac axis and the renal arteries. There was marked arterial sclerosis proximal to the coarctation. Wang,24 in 1950, reported the case of a 56-year-old man with congestive heart failure who had a coarctation 3 cm. above and at the level of the renal arteries. Doumber and co-workers25 described the case of a 49-year-old hypertensive patient with intermittent claudication. Aortography showed a stenosis at the level of the renal arteries with visualization of the right, but not the left artery. A left nephrectomy was performed without benefit. Goldzieher and associates26 described a 45-year-old woman with paroxysmal hypertension who was explored for possible pheochromocytoma, but a coarctation at the level of the renal arteries was found. Fisher and Corcoran27 recorded a case of a 14-year-old boy with blood pressure of 238/160 in the arm and 256/100 in the left leg. A preoperative aortogram was apparently normal. At laparotomy, the right kidney was smaller than the left and no pulse was palpable in the right renal artery. A right nephrectomy was performed. The patient died postoperatively, and postmortem examination showed a constriction of the aorta at the level of the renal arteries which had only slit-like orifices. The histologic picture was similar to that of congenital coarctation of the isthmus. In 1952, Albanese and Balia28 operated on a 12-year-old girl who had hypertension in the upper extremities, absent pulses in the abdominal aorta and femoral arteries, and a retrograde aortogram that showed no filling above the renal
arteries. At exploration, the aorta was hypoplastic from the diaphragm to the renal arteries with a markedly narrow area just above the renal arteries. This area was resected and a homograft was inserted, but the patient died postoperatively because of uremia and hypertensive encephalopathy. In 1950, Inmon and Pollock\textsuperscript{29} reported on the autopsy findings in a 32-year-old white woman who had a constriction of the aorta just below the renal arteries. A thrombus extended upward from this narrowed point occluding the left renal artery and impinging upon the orifice of the right renal artery. Ritchie and Douglas\textsuperscript{30} discussed the case of a 47-year-old man with intermittent claudication, blood pressure in the arm of 140/109, and no femoral pulses. At laparotomy, the aorta was found to terminate at the level of the renal vessels in a pulsatile cul-de-sac. Below this, an atrietic tube of 0.5 cm. passed down to bifurcate into 2 narrowed iliac vessels. A bilateral sympathectomy was performed with some improvement. Gerbasi and associates\textsuperscript{31} recorded a case in 1958, in which a homograft was successfully inserted to bypass a coarctation in the region of the celiac axis. Their patient was an 11-year-old girl with hypertension in the upper extremities and decreased femoral pulsations. An angiocardiogram was interpreted as showing a typical coarctation but at thoracotomy the coarctation was felt lower down. Postoperatively, a retrograde aortogram showed a coarctation 2 to 3 cm. above the renal arteries. At a second operation, the graft was successfully inserted.

Multiple coarctations are indeed rare lesions and, to our knowledge, there are reports of only 7 cases in the literature. In each case, the coarctation consisted of 2 areas of stenosis. In 1937, Benkwitz and Hunter\textsuperscript{32} reported the case a 67-year-old man with hypertension who died of pneumonia. No mention was made of the femoral pulses. At postmortem examination, 2 definite constrictions were found. The aorta became constricted at the beginning of the transverse arch and reduced to a diameter of 1 cm. at the site of the infantile isthmus. Two centimeters distal to the first coarctation, the descending aorta presented an annular constriction with almost complete obliteration of the lumen by diaphragm. The ligamentum arteriosum was located at the site of the second coarctation. In their article Benkwitz and Hunter discussed 3 previously described similar cases.\textsuperscript{33-35} In these cases the aorta was first constricted immediately beyond the origin of the left subclavian artery with a second area of stenosis at the level of the closed ligamentum arteriosum. Brock,\textsuperscript{36} in 1953, reported the case of a 17-year-old girl whose aortogram revealed what was apparently the usual type of coarctation. However, at thoracotomy a second area of stenosis was seen, 1 cm. above the main coarctation. The lower lesion was narrower than the upper. Between the 2 coarctations the aorta was thin and bulging. Both lesions were resected and a homograft was successfully inserted. No description was given of the microscopic pathology. In 1955, Cooley and DeBakey\textsuperscript{37} reported a 35-year-old man with a blood pressure of 260/160 in the left arm and no detectable pressure in the left leg. At operation, a constricting lesion, which began in the lower third of the descending thoracic aorta and extended a distance of 8 cm., was excised and replaced with a homograft. The constriction was surrounded by scarring and fibrosis. Because the leg pulses remained weak following surgery, retrograde aortography was performed. This showed a constriction in the proximal abdominal aorta just above the superior mesenteric artery and occluding the celiac artery. At a second operation this constriction, also atherosclerotic and calcified, was excised and replaced with a second homograft. A recent report of a double coarctation is that of Geronimo and associates.\textsuperscript{38} Their case was an 11-year-old girl with a blood pressure of 260/120 in the right arm, 160/130 in the left arm, and no femoral pulses. Previously the blood pressure in the left arm had been completely absent. He attributed the changes in pressure to occlusion and recanalization. An aortogram revealed a long constriction in the lower thoracic aorta and a second area of narrowing in the ab-
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dominant aorta above the superior mesenteric. Between the 2 constrictions, the aorta was dilated. Both segments were resected and successfully replaced with a homograft.

DISCUSSION

The work of Edwards and associates has led to the belief that the usual type of coarctation is due to a developmental defect of the media of the aorta. A localized medial thickening occurs as the primary lesion. In older patients a secondary intimal thickening may also occur. The microscopic findings in our case are perfectly consistent with the diagnosis of multiple congenital coarctations of the aorta. The occurrence of severe atherosclerotic changes and calcification proximal to an area of stenosis is well documented. Unfortunately, many of the cases of lower aortic stenotic lesions reported in the literature do not include a precise description of the histologic findings. The majority of the stenosing lesions in the descending thoracic aorta are of the long tapering variety. Their appearance has in general been considered by most authors as compatible with acquired inflammatory changes. The more localized lesions without evidence of inflammation have been thought of as congenital in origin.

Narrowing of the abdominal aorta may occur in any region, but commonly is either below or at the level of the renal arteries. These lesions for the most part, have been regarded as congenital in origin.

The 4 initially reported cases of double coarctation were similar. They consisted of constricted areas localized immediately beyond the origin of the left subclavian and at the level of the ligamentum. They were probably of congenital origin. Brock’s case also resembled these early reports. Cooley and DeBakey felt that in their case each of the constrictions was acquired. These lesions were arteriosclerotic and calcified and surrounded by scarring and fibrosis. They believed the etiology was a localized aortitis resulting in segmental arteriosclerosis with obliteration of the aortic lumen. In Geronimo’s case, the aorta was surrounded by dense inflammatory adhesions. The stenotic areas corresponded to deposits of granulation tissue in the media. The authors believed that the lesions were essentially an inflammatory destruction of the media.

DIAGNOSIS AND TREATMENT

The diagnosis of lower thoracic or abdominal coarctation should be suspected in any patient with relative hypertension in the upper extremities who, in addition, has evidence of increased collateral circulation over the lower chest, back, or abdomen. Murmurs, pulsations, or thrills in these regions should alert one to the possibility of an atypically located coarctation. In uncomplicated coarctations, intermittent claudication occurs in 8 per cent of the cases, but apparently is of greater incidence in abdominal coarctations, occurring in approximately one third of the cases. In typical coarctations, the aortic knob is small and may be absent. When the coarctation is in the lower thorax or abdomen, the aortic knob is normal and notching may be present only in the lowest ribs. If any of the above signs or symptoms are found, we consider it vital to delineate the exact site of the coarctation, with thoracic and abdominal aortograms. Intravenous angiography has resulted in misinterpretation of aortic lesions because of dilution of the dye. Particular attention should be paid to the length of the lesion and its relation to the renal arteries, for this will determine the operative procedure. Resection and grafting, bypass procedures, or even separate perfusions of the renal arteries might be necessary. It is not inconceivable that all 4 lesions in our case might have been successfully resected had we recognized them.

SUMMARY

Reports of multiple and infraducalectal coarctations are appearing more frequently in the literature as interest in the treatment of cardiovascular disease grows.

A case with 4 distinct coarctations is presented.
SUMMARIO IN INTERLINGUA

Reportos de coarctationes multiple e infraductal del aorta deveni de plus in plus frequente in le litteratura con le crescentia del interesse pro le tractamento de morbo cardiovascular.

Es presentate un case con 4 distincte coarctationes.

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ZAROFF, KREEL, SOBEL, BARONOFSKY


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