Pseudocoarctation and Mid-Arch Aortic Coarctation

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A 21-year-old woman was found to have a mid-arch aortic coarctation in combination with pseudocoarctation. The angiographic diagnosis was established by left atrial injection after transseptal puncture.

Since the first description of aortic coarctation in 1760, an array of clinical, radiologic, surgical and pathologic features have been recognized as typical of this common form of congenital heart disease. The coarctation usually occurs at or near the aortic isthmus. A less common form of aortic coarctation, located proximal to the left subclavian artery, has also been recognized. Twenty-five years ago, several authors described an entity characterized by deformity and mild narrowing of the aorta at the level of the isthmus with varying degrees of proximal dilatation and tortuosity. Controversy has existed over the etiology and significance of "pseudocoarctation," as this entity has become known. We describe an unusual form of coexistent proximal aortic coarctation and pseudocoarctation.

CASE REPORT

A 21-year-old Haitian woman was referred for evaluation of abnormal chest film findings. A heart murmur had been recognized when she was age three. Over the past several years, she had noted mild exertional dyspnea and calf pain. An aortic arch study by right brachial artery cutdown had been performed at another hospital, but had not provided adequate visualization of the aortic arch.

Physical examination revealed a normal body habitus. Blood pressure in the right arm was 126/80 mm Hg and in the left arm and calf was 90 by palpation. There was marked pulsation of the right carotid artery; the left carotid pulse was barely palpable. A left infraclavicular systolic bruit was heard. The heart, abdomen and extremities were normal. Femoral and pedal pulses were normal.

Routine laboratory data and electrocardiogram were normal. The chest film (Fig 1) revealed a widened superior mediastinum, extending both to the right and left of the manubrium, obscuring the usual location of the aortic knob. The lateral chest film showed a poorly defined density just behind the superior portion of the sternum.

Right and left heart catheterization were performed by percutaneous technique from the right groin. The study revealed normal intracardiac pressures and cardiac output. No intracardiac shunts were present. Blood pressure in the descending thoracic aorta was 93/80 mm Hg. An end hole catheter was passed through a tortuous segment of the distal aortic arch. A somewhat damped, but clearly higher pressure of 122/82 mm Hg was recorded at the mid-arch. The catheter could not be advanced beyond this point. No pressure gradient was found between the distal aortic arch and the distal thoracic aorta. The left atrium was entered by transseptal puncture and the catheter was advanced into the left ventricle. Simultaneous recording of left ventricular and descending aortic pressure revealed a 39 mm Hg pressure gradient between these two sites (Fig 2).

An aortic arch angiogram, obtained by left atrial injection of 60 ml of contrast medium (Renografin 76), (Fig 3)
demonstrated mild dilatation of the ascending aorta and the innominate artery. The sweep of the arch was unusually cephalad. Coarctation of the aortic arch was demonstrated at the origin of the left carotid artery which was noted to be severely hypoplastic. Distal to the coarctation was an "S" shaped, dilated segment of the aortic arch which involved the origin of the left subclavian artery. This segment had the radiologic appearance of pseudocoarctation of the aorta.

**DISCUSSION**

In 1940, Bayley and Holoubek reviewed the literature and stated that only 15 cases of aortic arch coarctation had been demonstrated. Several series derived from the medical and surgical literature suggest that aortic arch coarctation comprises 2.5 to 3.5 percent of all aortic coarctations. The majority of arch coarctations occur between the left carotid and the left subclavian arteries. Coarctation proximal to that site is considerably less common.

A variety of lesions may be responsible for lowered blood pressure in the left arm in patients with coarctation, including classic coarctation with atresia or stenosis of the left subclavian artery. A rare form of coarctation in which the right subclavian artery arises distal to an arch coarctation may present with normotension in all limbs and severe hypertension in the carotid arteries. Preoperative cardiac catheterization and angiography are appropriate in such atypical forms of coarctation.

A variety of approaches may be useful in the performance of these studies, including aortic study by the femoral, brachial or axillary techniques. This patient had previously had a right brachial artery cutdown and the retrograde arterial catheter could not be passed into the ascending aorta. The transseptal approach has been used widely to evaluate a variety of cardiac and vascular lesions including aortic dissection and is associated with a low incidence of complications in experienced hands. Left atrial angiography results in clearer visualization of left heart structures than does a pulmonary artery injection, and left atrial catheterization permits a thorough and accurate evaluation of hemodynamic parameters. To those experienced in its use, it represents a very useful alternative technique.

Beginning with several case reports in 1951 about 100 cases of pseudocoarctation of the aorta have been reported. Additional descriptive terms have included subclinical coarctation and kinking or buckling of the aorta. Attention was first called to this abnormality by the radiologic appearance of a superior mediastinal mass. This appearance is produced by tortuosity and dilatation of the distal aortic arch, related to mild narrowing or kinking of the aorta at the level of the isthmus.

Edmunds and colleagues analyzed the size of the normal aorta proximal to, at the site of, and distal to the aortic isthmus. He found that in normal individuals the aorta at the isthmus has at least 80 percent of the cross sectional area of the adjacent aorta. In clinically significant coarctation, the cross sectional area of the aorta is reduced by at least 60 percent. Between these two ranges an unknown number of individuals may have a subclinical form of coarctation.

While the majority of patients with pseudocoarctation do not have an intra-aortic pressure gradient, a number of reports have demonstrated small gradients. In one case report a gradient of more than 50 mm Hg was found. Several of these cases were reported as examples of combined coarctation and pseudocoarctation, but no angiographic, surgical or pathologic evidence of true coarctation was presented. Pressure gradients across the "kinked" area of a pseudocoarctation are now a well documented feature in some cases. The present patient is remarkable for the association of clearly documented coarctation and pseudocoarctation.

The possibility that these two malformations of the aorta may be etiologically related should be considered. The presence of pressure gradients in pseudocoarctation and the absence of significant gradients in some cases of classic coarctation has led to speculation that the distinc-

**Figure 3a (left).** Aortogram in left anterior oblique projection. The uppermost arrow indicates the atretic left carotid artery. The lowest arrow indicates the tortuous segment of the distal aortic arch. The thin arrow indicates the site of the mid-arch coarctation. **Figure 3b.** (right). Line drawing of angiogram in Figure 3a. Rt = right; Lt = left; Innom Art = Innomin

Artery; Asc = ascending; Desc = descending.
tion between the two entities may be artificial. Smyth and Edwards\(^5\) have presented this argument, citing the presence of a localized ridge of medial tissue projecting into the aortic lumen in classic coarctation and in several cases of pseudocoarctation which they examined. However, surgical and pathologic material from other cases has not confirmed the universal presence of such a ridge in pseudocoarctation.\(^6\) Dungan et al\(^7\) reported the absence of any intraluminal obstructing lesion in several cases of pseudocoarctation in which pressure gradients were identified. The gradient is presumably due to the kinking of the aorta; amelioration of pressure gradients after surgical relief of the kinking has been reported.\(^8\)

Pseudocoarctation appears to be a congenital, rather than an acquired anomaly. Its appearance in infants\(^9\) and its association with other forms of congenital heart disease strengthen this conclusion. Souders and associates\(^2\) in their original description, suggested that traction on the aorta by an abnormally short ductus might be the cause of pseudocoarctation. Pattinson and Grainger\(^17\) postulated that failure of normal embryologic compression of the third to seventh segments results in pseudocoarctation. Lavin et al\(^11\) proposed that failure of the distal right aortic arch to atrophy and its subsequent incorporation into the distal left arch might be the embryologic basis of pseudocoarctation.

The unusually cephalad course of the aortic arch, extending both to the left and right of the sternum, suggests that the third, rather than the fourth embryologic arch may have persisted in this patient. The coincidence of this finding, together with the mid-arch coarctation and the pseudocoarctation itself, supports the contention that pseudocoarctation represents a congenital anomaly. It is possible that the tortuosity and dilatation of the aorta distal to the coarctation might merely represent post-stenotic dilatation. However, this degree of post-stenotic change is rarely observed in mid-arch coarctation. The relationship between coarctation and pseudocoarctation remains conjectural.

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Interstitial Pneumonitis Complicating Rheumatoid Arthritis

Sustained Remission with Azathioprine Therapy*\(^8\)

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A patient with classic rheumatoid arthritis developed biopsy-proven diffuse interstitial pulmonary fibrosis and ventilatory insufficiency which appeared to be irreversible. The administration of azathioprine coincided with significant immediate improvement in pulmonary function and clinical status. During five years of continuous azathioprine therapy, progressive improvement in lung function has been accompanied by marked deterioration of the rheumatoid joint disease, suggesting that the pulmonary and joint lesions of rheumatoid disease may not be mediated by the same pathways.

Although arthritis is the most common manifestation of rheumatoid disease, involvement of other organ systems has been described. Ellman and Ball\(^1\) first noted diffuse interstitial pneumonitis with fibrosis at autopsy of two rheumatoid patients. While the association of interstitial pneumonitis with fibrosis to rheumatoid dis-

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