Pseudocoarctation of Aorta Associated with Aneurysm Formation*

S. Bahabozorgui, M.D., Robert G. Bernstein, M.D., and R.W.M. Frater, M.D.

We report a patient with a large, thin-walled saccular aneurysm below the kinked area of the aorta. This was successfully resected. Review of the literature discloses a similar case that ruptured. Pseudocoarctation of the aorta cannot, therefore, be regarded as entirely benign: it may produce hemodynamic conditions for development of aneurysm. Careful follow-up and appropriate surgical treatment is recommended for all cases.

Pseudocoarctation of the aorta has been recognized as an anatomic entity which produces a characteristic radiologic deformity. Although there has been considerable emphasis in the literature concerning its benign outcome, in recent years several cases of aneurysm formation have been reported and Gay and Young were forced to reappraise this condition after one of their patients with pseudocoarctation died from rupture of the aorta. We recently operated on a young asymptomatic patient with pseudocoarctation and aneurysm formation. The purpose of this paper is to reemphasize the potential complication of this disease and its further evaluation.

Case Report

A 20-year-old white woman was admitted to the Hospital

*From the Departments of Surgery and Radiology, Albert Einstein College of Medicine, New York City.

FIGURE 1. PA chest film on admission shows an elongated aortic arch with aneurysmal dilatation of the descending aorta below a kinked segment (+). Rib notching is not present nor is there cardiomegaly.

of the Albert Einstein College of Medicine for investigation of a lesion found in a routine chest x-ray examination at the Immigration Service. She was asymptomatic and denied any significant past medical history. Chest x-ray film had revealed a mass in the left superior mediastinum adjacent to the aortic shadow (Fig 1 and 2). The blood pressure was 110/70 and equal in all extremities. There was a questionable bruit over the right carotid artery. Examination of the chest gave

FIGURE 2. AP film during axillary aortography clearly shows an elongated aortic arch with kinking and aneurysm formation. Collateral circulation is not present. The aortic valves are normal.
PSEUDOCOARCTION OF AORTA

Figure 3. LAO film shows the filling defect of the internal narrowing that was seen at surgery at the level of the kinked segment (>). The aneurysm is immediately below this.

negative results. A grade II/VI systolic murmur was heard at the base of the heart with radiation to the neck. Physical examination was otherwise negative. All laboratory values were within normal limits. The electrocardiogram was normal. An axillary aortogram revealed an elongated aortic arch, with kinking and narrowing of the aorta distal to the origin of the left subclavian artery and aneurysm formation distal to this narrowed area. The aneurysm measured 8 by 6.5 cm. There was no evidence of rib notching or collateral circulation. The descending aorta was normal (Fig 3).

On October 23, 1969, the patient underwent surgery. Partial cardiopulmonary bypass was employed. The aortic arch was found to be elongated and high in the mediastinum. Below the left subclavian artery, at the site of the ligamentum, the aorta turned sharply toward the midline and as sharply returned to its normal course. At this point, there was moderate narrowing which, from the inside, had an appearance not unlike some coarctations, except that it was so much wider and that the tissues were not at all thickened. Distal to this narrowing and buckling, there was a saccular aneurysm 8 cm in diameter with a thin wall. The aneurysm was resected and a woven Teflon prosthesis was used for replacement. The postoperative course was uneventful.

DISCUSSION

The clinical and radiographic features have been well described and the differential diagnosis from true coarctation, aneurysm and mediastinal tumor extensively discussed. It is important to note that hemodynamically there is an absent or a very small gradient across this deformed area, indicating that the area of the lumen of the stenotic segment is greater than the critical 25 to 35 percent of the cross-sectional area of the proximal aorta that is needed to produce a gradient. This anomaly can usually be differentiated from "true" coarctation of the thoracic aorta by the absence of collateral circulation, systemic hypertension, femoral pulse lag, and a pressure gradient across the "kinked" segment, and the presence of a conspicuous aortic knob. It has also been suggested that this is an abortive form of true coarctation.

Although all emphasis in the past has been on the benign nature of the lesion and accurate diagnosis has been stressed to avoid unnecessary thoracotomy, several cases of aneurysm formation and surgical resection have now been reported. The precise incidence of aneurysm formation is not yet clear, but the pathogenesis of this dilated and thin-walled segment of aorta is presumably related to turbulent flow beyond the kink. Despite the absence of gradients, the presence of systolic murmurs in most patients attests to the probability of turbulence. With conversion of forward energy to lateral pressure, the distal aorta presumably is stretched and this in turn may aggravate the kink. Mycotic aneurysm, although known with true coarctation, has not been reported as a complication of pseudocoarctation. Congenital anomalies such as aortic stenosis, subaortic stenosis, patent ductus arteriosus, ventricular septal defect, single ventricle, atrial septal defect and anomalies of the branches off the aortic arch associated with pseudocoarctation have been reported. Treatment of this lesion in an asymptomatic patient with or without associated anomalies or complications has been conservative in the past. However, close follow-up of a known patient is very important. It is quite possible that surgical treatment will be desirable in the future after more information is obtained about the natural history of the disease. It would seem reasonable that complicating aneurysm formation should be treated aggressively.

REFERENCES


Reprint requests: Dr. R. W. M. Frater, Department of Surgery, Albert Einstein College of Medicine, Bronx 10461