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The Surgical Treatment of Pseudocoarctation: A Case Report

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Abstract

A 35-year-old female with pseudocoarctation underwent an operation in which total resection of the buckled segment and subsequent Dacron graft replacement were performed. Intimal surface of the resected segment had folds that protruded into the lumen, but other areas were extremely thin and slightly calcified, indicating a great possibility of rupture. Excluding an asymptomatic patients with minimal deformity and thick aortic wall, surgical treatment should be performed to prevent rupturing.

Introduction

Pseudocoarctation was first described independently by DOTTER and STEINBERG[3], ROBB[8] SOUDERS et al.[10] in 1951. Other than pseudocoarctation, subclinical coarctation[4], and kinking or buckling of the aortic arch are used in the terminology of this lesion. According to LAVIN[6], this anomaly can be defined as an elongation and kinking of the aortic isthmus which differs from true coarctation primarily by the absence of aortic pressure gradients and collateral circulation.

Most patients with pseudocoarctation have no indication of surgical treatment. In some

Key words: Pseudocoarctation of the aorta, Aneurysmal dilatation, Transient bypass, Rupture, Histological study
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patients, however, rupture occurs due to pseudocoarctation. We encountered a patient with pseudocoarctation that posed a great risk of rupturing; the lesion was resected and end-to-end anastomosis was performed. The clinical course of this patient is reported.

Patient

A 35-year-old female was first found to have a murmur at the age of 12 years. Since then her physical activity was limited; however, she complained of shoulder stiffness, dizziness, tinnitus, and intermittent claudication. At the age of 17 years, she experienced an episode of loss of consciousness with throbbing headache and nausea. She was admitted to a hospital for 40 days, where she was diagnosed as having aortic arch syndrome on the basis of angiographic findings.

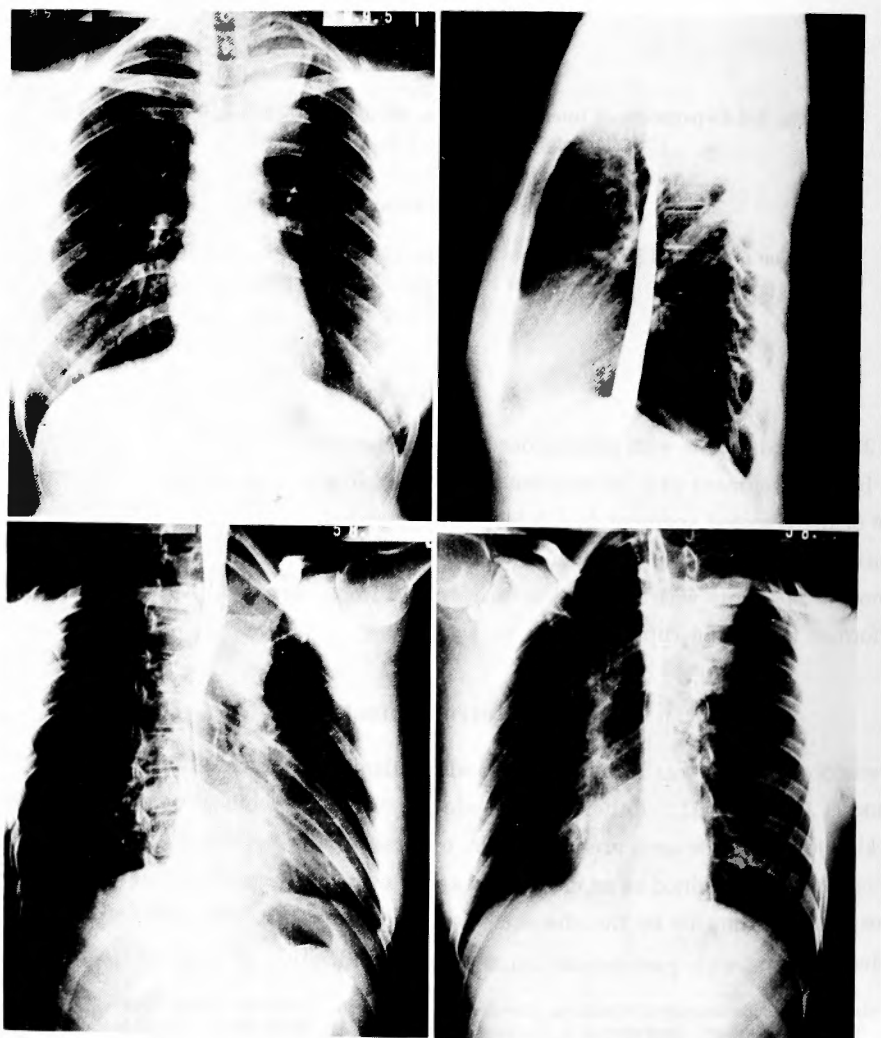

Fig. 1. Preoperative roentgenography showing an abnormal shadow in the left superior mediastinum.

Moreover, a catheter, inserted from the right brachial artery and right femoral artery, could not be advanced into the ascending aorta. These findings suggested the presence of stenosis in the aortic arch and torsion of the subclavian artery; a more precise diagnosis was not made. At that

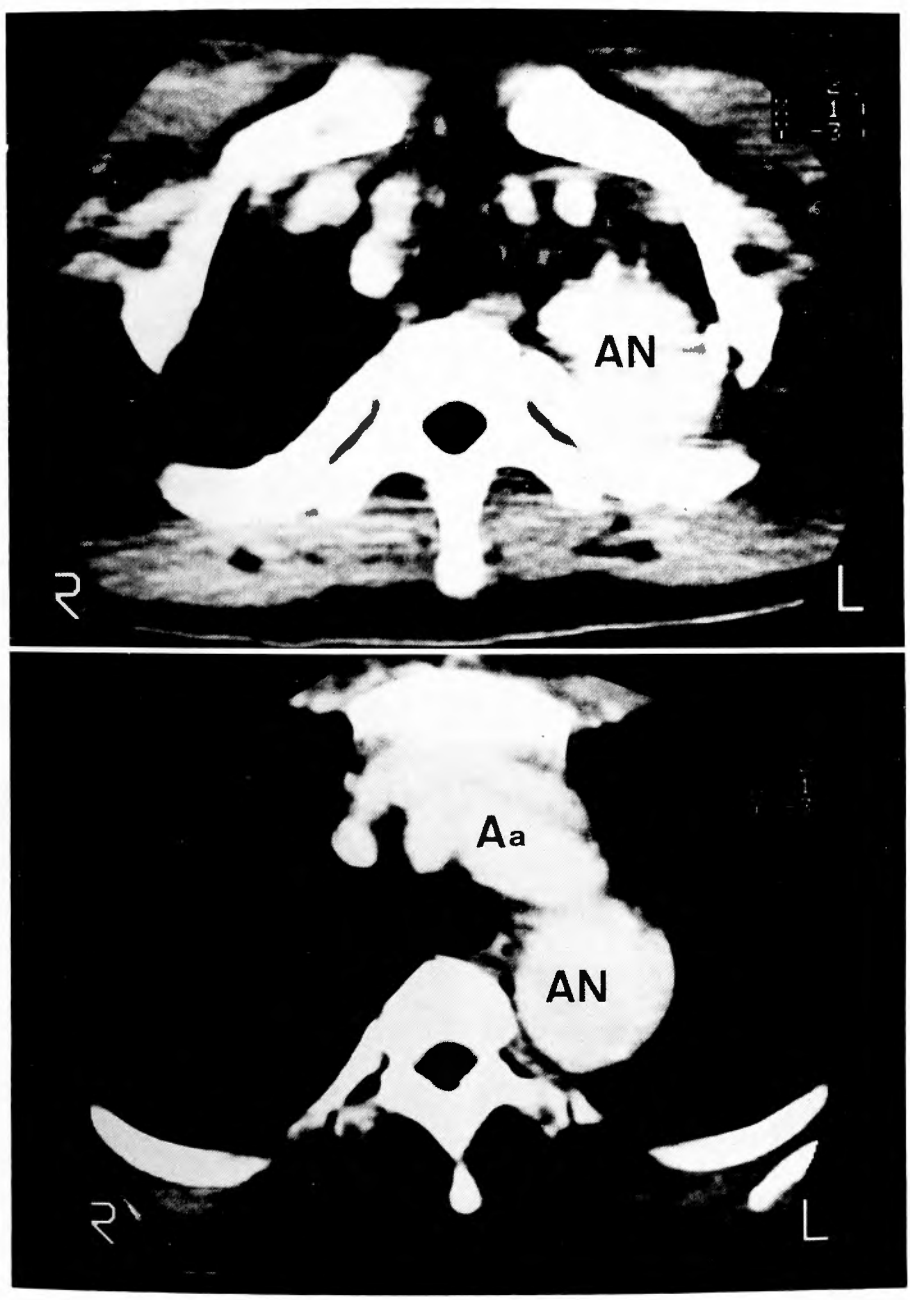

Fig. 2. Preoperative CT scan
AN: aneurysmal dilatation of the descending aorta
Aa : aortic arch

time, hypertension in the upper half of the body, a blood pressure gradient of 30 mmHg between the right and left brachial arteries, and difficulty in measuring blood pressure in the lower extremities were noted. Being asymptomatic, she was discharged without treatment. At the age of 33 years she consulted a local hospital in order to obtain detailed information concerning her disorder, and was subsequently reffected to our hospital on March 18, 1981. Various examinations disclosed a pseudocoarctation in the proximal descending aorta. CT scan revealed a very thin aortic wall with a great possibility of rupture. There was no history of thoracic trauma or chest pain.

The patient was 155 cm tall and weighed 52.8 kg. Blood pressure was 110/64 mmHg in the right brachial artery, 110/70 mmHg in the left brachial artery, 100/62 mmHg in the right leg and 94/64 mmHg in the left leg. A grade 3–4/6 harsh systolic murmur was present over both supraclavicular regions, radiating widely to the anterior chest and the back. Thrill was not palpable. Chest roentgenography revealed a left superior mediastinal mass; there was no rib notching (Fig. 1). Electrocardiography demonstrated no evidence of left ventricular hypertrophy. CT scan showed coexisting stenosis and aneurysmal dilatation in the proximal descending aorta, the wall of which was slightly calcified (Fig. 2).

Cardiac catheterization was performed on May 13, 1983. Pressures of each chamber were normal. Cardiac output was 4.03 l/min, and cardiac index 2.71 l/min/M^2. Angiography revealed coexisting stenosis and aneurysmal dilatation in the proximal descending aorta, coiling of the right subclavian artery and occlusion of the left vertebral artery (Fig. 3). These findings corresponded to those of pseudocoarctation. Laboratory findings were within normal limits.

Operation was performed with the use of surface hypothermia on September 12, 1983.

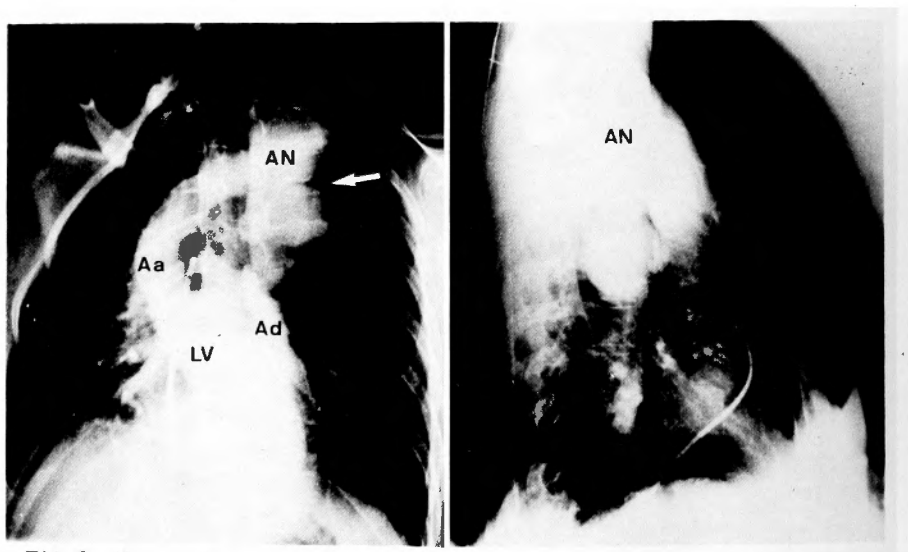

Fig. 3. Preoperative aortography showing coexisting stenosis and aneurysmal dilatation in the proximal descending aorta.
Aa: ascending aorta, Ad: descending aorta, LV: left ventricle, AN: aneurysmal dilatation of the descending aorta (arrow)

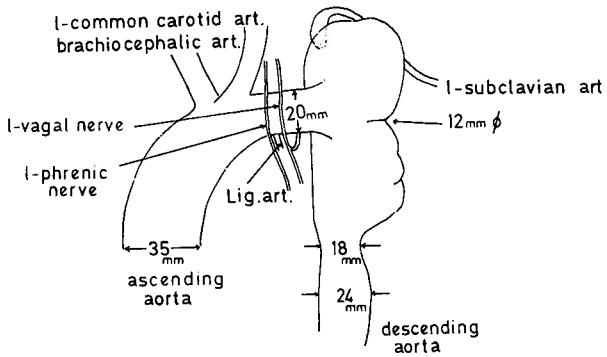

Fig. 4. Operative findings.

Thoracotomy was performed in the left 4th intercostal space, extending to the right 4th intercostal space adding a horizontal division of the sternum. This incision permitted a wide operative field of the ascending aorta, aortic arch and descending aorta. No adhesion between the lung and aneurysm was found. The aneurysm consisted of two dilated sections of the descending aorta (Fig. 4). A portion of the lower aneurysmal dilatation was yellowish, and its wall was very thin. Ligamentum arteriosum was atypically situated between the left common carotid artery and aneurysmal dilatation, from which the left subclavian artery arose. The ligamentum was double ligated and divided. Using a woven Dacron graft (70 cm in length, 12 mm in diameter) pre-clotted with fibrin glue, a transient bypass was performed between the ascending aorta and descending aorta. The aortic arch distal to the left common carotid artery, and the descending aorta distal to the aneurysm were taped and corss-clamped. Carefully avoiding damage to the vagal, recurrent and phrenic nerves, the aneurysmal dilatation of the descending aorta was

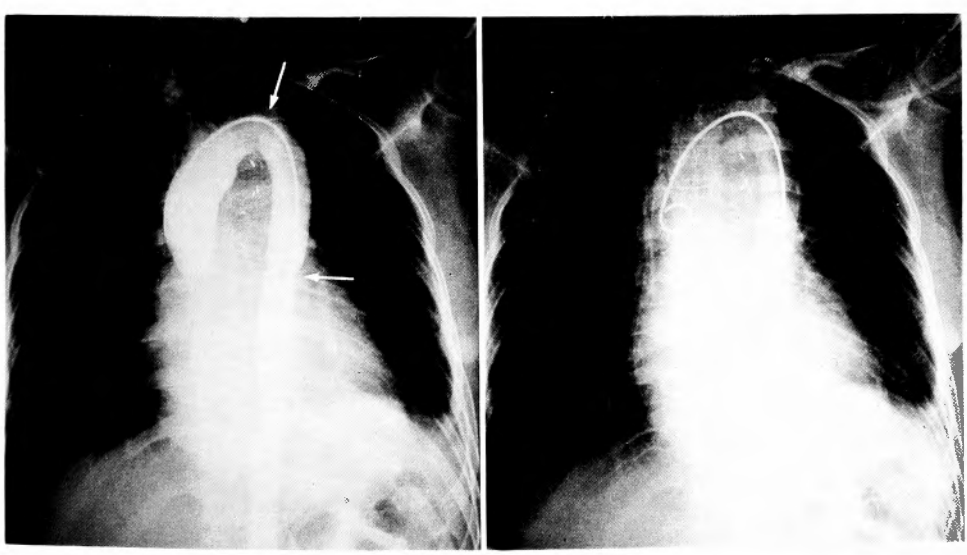

Fig. 5. Postoperative aortography showing the graft between the arrows without pressure gradient.

resected. During this procedure, the intercostal arteries, the sizes of which were normal, were ligated or clamped using silver clips. The left subclavian artery arose from the upper aneurysmal dilatation of the aorta. A second Dacron graft (7 cm in length, 20 mm in diameter) was placed

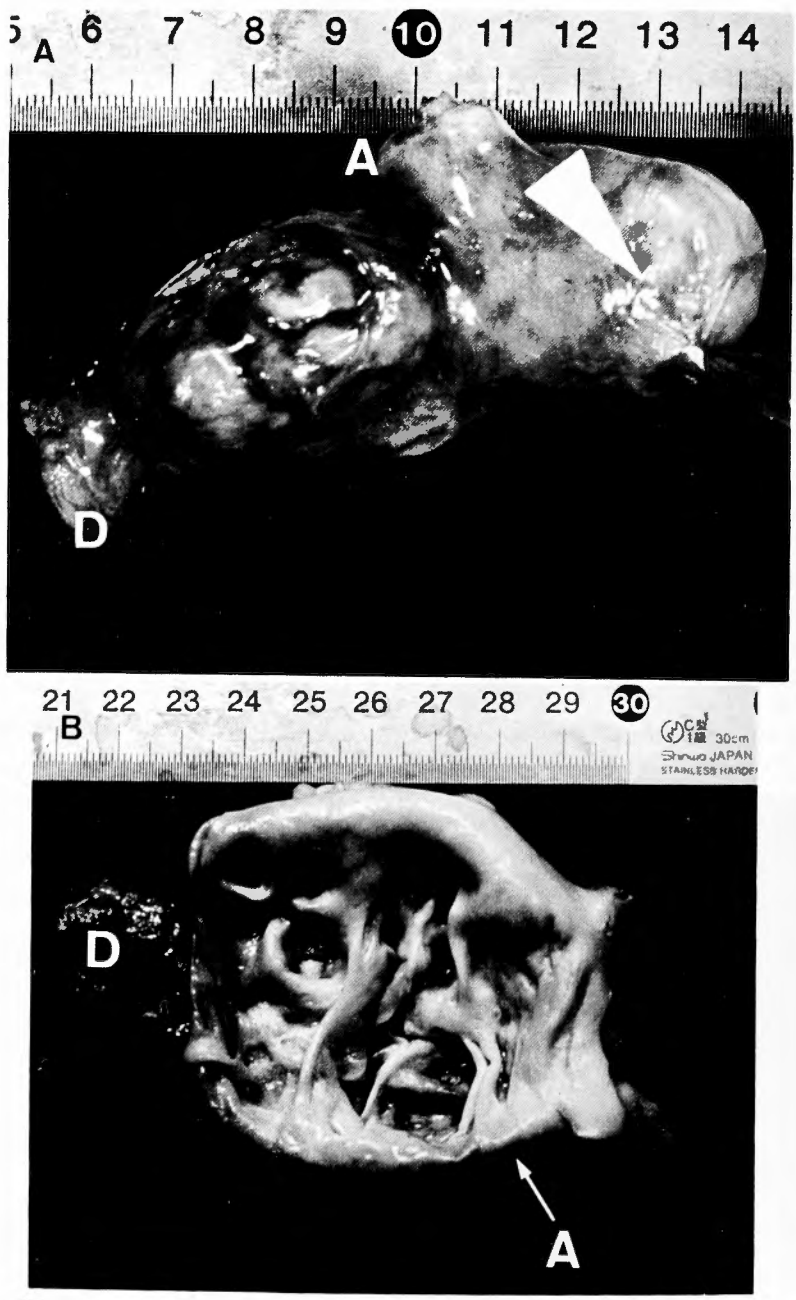

Fig. 6. Surgical specimen
A) gross appearance
B) intimal surface
A: aortic arch, D: descending aorta, arrow showing calcified portion.

between the aortic arch and descending aorta. The left subclavian artery was anastomosed to the graft, interposing a Gore-tex graft (6 mm in diameter). Both anastomoses were wrapped by the woven Dacron graft used as a transient bypass.

In the postoperative period, left pleural effusion was noted, but could not be aspirated per-

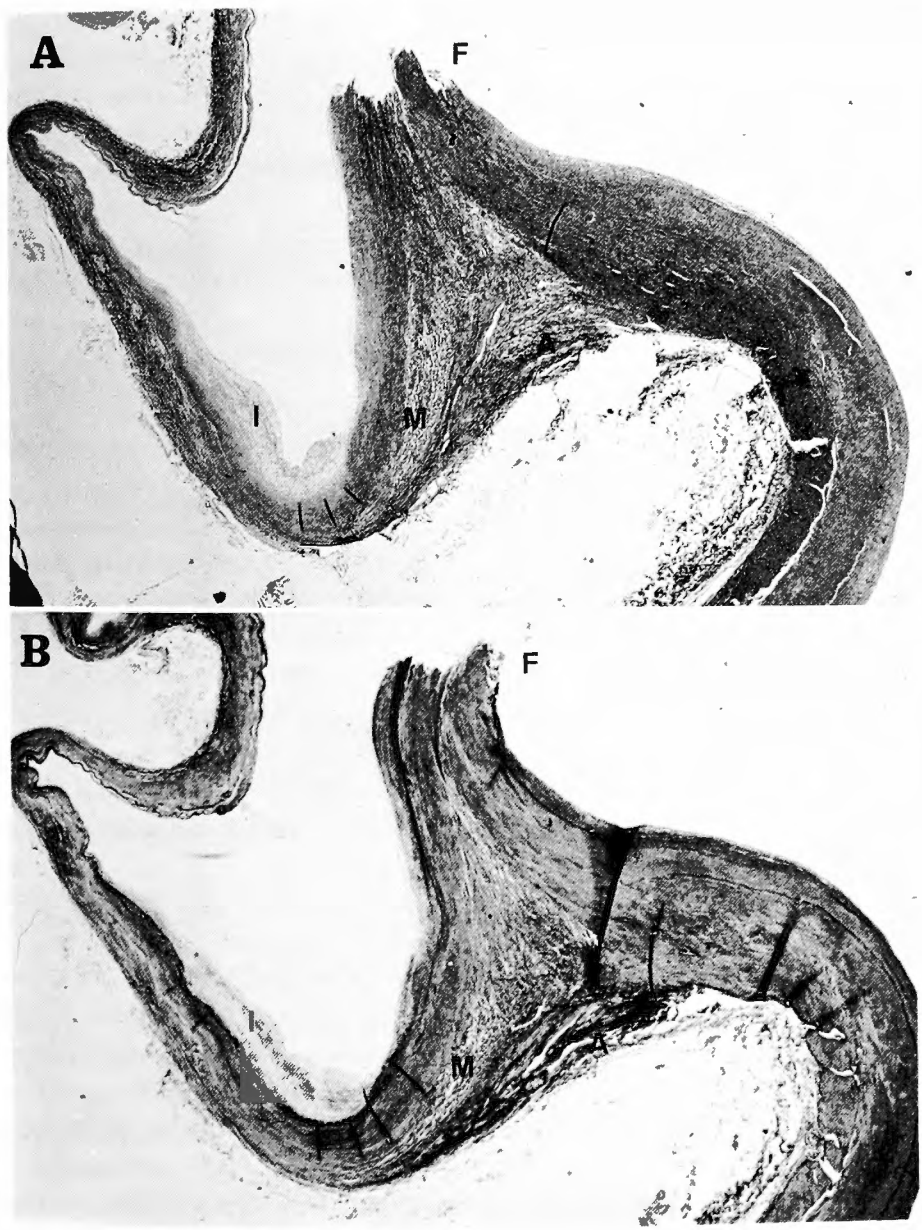

Fig. 7. Photomicrographs of the aortic wall with pseudocoarctation
A) Hematoxylin-Eosin stain (×4)
B) Van Gieson stain (×4)
F: fold, I: intima, M: media, A: adventitia

cutaneously because of its gelatin-like character. However, three weeks later, the effusion disappeared spontaneously. Pulse of the left brachial artery was weaker than the right.

Postoperative cardiac catheterization and angiography revealed neither pressure gradient across the graft nor stenosis; the root of the left subclavian artery was occluded completely, but the visualization of the distal portion due to collateral circulation was slightly delayed (Fig. 5).

Surgical specimen: The resected segment of the aorta was 8.7 cm long. The proximal end was 22 mm in diameter, and the distal end 18 mm (Fig. 6). At the midportion, the outside diameter was constricted to 12 mm. The intimal surface of the involved portion had folds protruding into the lumen. Some small areas in the anterior wall of the lower dilated segment were very thin. Microscopically, the irregular thinning was remarkably found with disappearance of normal aortic wall structure (Fig. 7). The major characteristics of this lesion are coexistence of hypertrophy and atrophy in the aortic wall. In the center of the large fold the media and adventitia appeared "T"-shape; it is speculated that one side of the aortic wall enlarged and subsequently collapsed, with the two folded portions adhering to each other. Calcification, myxomatous degenerative change, and rupture of elastic fibers were also found in some portions.

Discussion

Pseudocoarctation, subclinical coarctation, and buckling or kinking of the aorta have been terms used to describe a roentgenologic deformity of the aorta similar to that in clinical coarctation but in which no obstruction is demonstrable[7,9,12]. GAY[5] listed the criteria for diagnosing pseudocoarctation as follows: 1) an abnormal postero-anterior chest roentgenogram (generally a left superior mediastinal density), 2) no, or small (less than 25 mmHg), pressure differential between arms and legs, 3) no evidence of increased collateral circulation (rib notching), and 4) a definitive aortogram. Therefore, this deformity of the aorta had been considered by many authors as a "benign clinical condition", warranting no specific therapy.[12] GAY[5], however, reported a 61-year-old patient who died of rupture of a dissecting aneurysm arising immediately distal to the pseudocoarctation and emphasized that symptomatic patients and those with severe aortic deformity, as demonstrated by aortography, should undergo an operation.

As seen in our case, in some areas folds of the aortic wall protruded into the lumen while in other areas extensive thinness or calcification was found. This latter site of definite weakness had a great possibility of rupturing. In addition, the discontinuity from this involved lesion to the normal aortic wall might be the cause of the dissecting aneurysm arising distal to the pseudocoarctation. The coexistence of the hypertrophic portion which formed a fold and the atrophic portion of the wall which was thin and convex, as viewed from the outside, is thought to be due to congenital fragility of the aortic wall. This characteristic can not be reasonably explained by embryologic development of the aortic arch[2,6]. Pseudocoarctation with pressure gradient less than 25 mmHg probably does not produce this lesion, but surely plays a role in its progression. Development of atherosclerotic changes may occur due to eddies that presumably exist in the kinked region of this abnormally shaped aorta[4]. In addition, the degenerative changes of senescence or hypertension increase the possibility of rupture.

Thus, surgical treatment in some patients is of a preventive nature in the absence of definitive indications. However, in other patients, surgical intervention is indicated as follows: 1) a great possibility of rupture resulting from thinness and degenerative change of the aneurysmal aortic wall, 2) more than 30 mmHg pressure differential between arms and legs, 3) co-existing coarctation[1], and 4) the presence of other associated diseases[11].

The operation generally consists of resection of the buckled segment along with dilated segments of the aorta; graft replacement is usually necessary. Occasionally, reconstruction of the left subclavian artery is required. A transient bypass between the ascending aorta and lower descending aorta does not necessitate cardiopulmonary bypass or double operative incisions. The operation is safe and easy because of the absence of adhesion between the lung and pseudocoarctation, no increased collateral circulation and the limited site involved.

An asymptomatic patients with minimal aortic deformity and thick aortic wall should be followed with periodical examinations such as roentgenography, CT scan and ultrasonic cardiography.

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和文抄録

Pseudocoarctation の外科治療

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35歳，女性，仮性大動脈縮窄症に対し，動脈瘤様拡大を来たした部分を全摘し，Dacron graft で置換した．摘出標本の内膜面には，大小種々の襞形成と，その間に存在する小さな瘤状形成が多数認められた．従来，仮性大動脈縮窄症に対し，外科手術の必要性はないと考えられて来たが 本例のように 大動脈壁が菲薄化し 破裂の危険が大なる場合 積極的に外科治療を行うべきである．