Subaortic Stenosis Caused by Two Discrete Membranes*

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A case of subaortic stenosis caused by two discrete membranes is presented. Following demonstration of the two distinct sites of obstruction by intraventricular pressure gradients and angiocardiogram, the two membranes were excised through a transaortic approach. The importance of being aware of the occurrence of this rare anomaly is stressed, and its surgical implications are discussed.

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Figure 1. Composite diagram of catheter withdrawal pressures from apex of left ventricle to ascending aorta, showing two distinct gradients in ventricular cavity and no gradient across aortic valve.

Fixed obstruction of the left ventricular outflow tract accounts for 8 to 10 percent of all cases of congenital aortic stenosis, which, in turn, represents approximately 5.5 percent of all congenital cardiac defects. The fixed obstruction has been reported to be usually due to a discrete subvalvular membrane or, less commonly, to a fibrous tunnel involving the entire left ventricular outflow tract.

We have observed a patient in whom fixed obstruction of the left ventricular outflow tract resulted from two distinct membranes, an anatomical situation that, to our knowledge, has not been previously described. The presentation of this case and the discussion of its diagnostic and surgical features are the objects of the present report.

Case Report

A 12-year-old boy was admitted with a history of fatigue and dyspnea on moderate exertion. A heart murmur was discovered when the patient was three years old; at age five years, he underwent cardiac catheterization in Rome, and a diagnosis of idiopathic hypertrophic subaortic stenosis was made. Since that time, the patient's activities have been restricted, and he was treated with propranolol hydrochloride (Inderal) since age ten without improvement.

The patient's blood pressure was 110/70 mm Hg. A systolic thrill was present maximally at the lower left sternal border and radiated over the precordium and the supra-sternal notch.

A chest roentgenogram showed cardiomegaly with left atrial and left ventricular enlargement and a prominent ascending aorta. An electrocardiogram disclosed left ventricular hypertrophy and strain pattern.

Cardiac catheterization showed the pressure at the apex of the left ventricle to be 210/18 mm Hg. The catheter pullback from the apex to the aortic valve (Fig 1) revealed two intracavitary pressure gradients, one of 75 mm Hg in the midportion of the left ventricle and the second of 40 mm Hg at the outflow-tract level. No gradient was recorded across the aortic valve.

The angiocardiogram (Fig. 2) revealed a large ventricular chamber with two discrete sites of obstruction, one immediately below the aortic valve and the second approximately...
1.5 cm lower, in contiguity with the anterior leaflet of the mitral valve. The aortic valve appeared to be normal.

At surgery, the normal appearing aortic leaflets were gently retracted; a crescent-shaped, thin membranous structure (Fig 3A) located immediately subjacent to the aortic valve came into view and was excised circumferentially. With the diaphragm immediately subjacent to the aortic valve removed, the ventricular cavity was inspected through the aortotomy for the second site of obstruction. This was apparent only after a thorough exploration, stimulated by the findings of cardiac catheterization (Fig 1 and 2). It appeared as a relatively thick, crescent-shaped fibrous diaphragm (Fig 3B) located approximately 2 cm caudal to the other membranous structure and attached in part to the septal leaflet of the mitral valve. The membrane was excised by sharp dissection; a thin rim of fibrous tissue was left attached to the mitral valve to avoid leaflet injury. At cessation of bypass, pressure measurements were obtained, and there was a residual gradient of 20 mm Hg between the left ventricle and the aorta at the level of the lower obstruction. Recovery was uneventful, and the patient remains asymptomatic one year after surgery.

Discussion

Two distinct types of fixed, discrete obstruction of the left ventricular outflow tract have been recognized.6,7 The first type is a membranous diaphragm located at the cranial portion of the left ventricular outflow tract immediately beneath the aortic cusps. The second type is usually described as a thick fibrous ring located more caudally and attached in part to the septal leaflet of the mitral valve. A moderate muscular or fibromuscular obstruction of the left ventricular outflow tract is frequently associated with this latter type of lesion. The extreme form of this malformation is represented by the left ventricular tunnel.8-10

In the case reported herein, there were two distinct obstructions formed by separate crescent-shaped diaphragms (Fig 3). The first was located immediately below the aortic cusps; the second was located caudally in close relationship with the septal leaflet of the mitral valve. Thus, in the case, both types of fixed, discrete obstruction of the left ventricular outflow tract that have been described were present.

Deutsch et al8 have classified a malformation in which there are multiple sites of obstruction as type 3 of discrete subaortic stenosis; however, this entity differs substantially from our case both in the anatomic structure and in its angiographic appearance.

Although both the pressure curves (Fig 1) and the angiogram (Fig 2) in our patient were suggestive of the diagnosis and made us aware before surgery of the existence of two separate, discrete sites of obstruction, visualization of the lower membrane after excision of the one immediately subvalvular was somewhat difficult; it may then be inferred that in the absence of a clear preoperative demonstration, a second obstruction might remain unrecognized at surgery.

The question of recurrence of discrete subaortic stenosis after surgical excision has been raised.1 Although this possibility may indeed be a real one, presumably with regard to any muscular or fibromuscular component of the lesion, the finding of an intact fibrous diaphragm at a second operation should, in view of our experience, raise the suspicion that there might have been a second membrane which went unrecognized at the first surgery. Therefore, the importance of a meticulous inspection of the subaortic area, supplemented by intraoperative simultaneous measurement of the left ventricular and systemic pressures, cannot be overemphasized.

We believe that in this case the lower obstruction was similar to those described as type 2 by Kelly et al,7 in which, in addition to a thick fibrous ring, moderate fibromuscular obstruction is present. It is known that in

Figures 2 and 3. Ventriculogram in right anterior oblique projection. Two discrete obstructions are visualized (arrows); one is immediately subjacent to aortic valve, and other is approximately 1.5 cm more caudal.

Figures 3. Surgical specimens of the two membranous subaortic obstructions that were removed. A, Thin subvalvular diaphragm; and B, Fibrous ridge partially attached to anterior leaflet of mitral valve.
such cases, surgical relief of the lesion often may not be complete, and late results are not as satisfactory as those obtained after excision of the thinner and more pliable obstructions located immediately subvalvularly. It is plausible, however, that the mild residual gradient demonstrated in our patient may undergo progressive resolution; the possibility of this course of events previously has been documented repeatedly in serial studies.*

REFERENCES


Resolution of Pulmonary Lymphangitic Carcinoma of the Breast*

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Pulmonary lymphangitic carcinomatosis, a grave complication of malignant neoplasms, usually progresses rapidly and is fatal despite all modes of therapy. We recently observed complete resolution of pulmonary lymphangitic involvement from an adenocarcinoma of the breast following both chemotherapy and bilateral oophorectomy. The patient's chest radiograph has remained clear for 1½ years.

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CHEST, 69: 1, JANUARY, 1976

Complete resolution of pulmonary involvement in metastatic breast carcinoma is unusual. A recent report documented a patient with disappearance of metastatic pulmonary nodules after bilateral oophorectomy. We examined a patient in whom there has been complete radiographic resolution of pulmonary lymphangitic involvement from an adenocarcinoma of the breast.

Pulmonary lymphangitic carcinomatosis, a diffuse neoplastic permeation of pulmonary and pleural lymphatics, is a grave complication of malignant neoplasms. The radiographic and pathologic characteristics of lymphangitic spread of tumor to the lungs have been well described. Although dyspnea may antecede demonstrable radiographic changes, characteristic abnormalities are usually present on chest radiographs at the time of diagnosis. Despite all modes of therapy, relentless progression until death is usually seen radiographically and clinically.

CASE REPORT

A 39-year-old woman was admitted to the University of California Medical Center for shortness of breath and a nonproductive cough of one month's duration. She had been in excellent health until five years previously when, in the seventh month of her fifth pregnancy, she noted a small lump in the upper outer quadrant of her left breast. It was biopsied and found to be benign. Three months later, the lump recurred. Biopsy revealed adenocarcinoma, and a left radical mastectomy was performed. There was no evidence of axillary nodal involvement at the time of the operation.

Seven months before admission, the patient began to experience intermittent low-back pain. Investigation at that time, including radiographs of the lumbosacral spine and an 18fluorine bone scan, revealed no abnormalities. Chest radiographs showed patchy nodular and linear densities of the lower lobe of her right lung.

FIGURE 1. Posteroanterior chest radiograph demonstrating ill-defined density involving both lower lobes with both linear and nodular components. Septal lines are present. Left radical mastectomy was performed previously.