Supra-valvular Stenosis of the Aorta *

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A STENOSING LESION of the aorta situated just above the aortic valve has been considered a distinct rarity and has been given relatively little attention. Archer [1] in 1878 was the first to mention an elastic band stretching across the origin of the aorta. Several similar reports followed, describing thin strands of tissue in the ascending aorta, apparently without hemodynamic significance. Mencarelli [8] in 1930 was the first to report a stenosing lesion of the aorta situated above the valve, and also coined the term: supravalvular stenosis of aorta.

With the recent advances in cardiovascular surgery, more aortic valves are being explored under direct vision and this lesion is likely to be encountered at the operating table, and possibly diagnosed preoperatively, with increasing frequency. With this in mind, further clarification of the clinical and anatomic picture, the natural history and the surgical possibilities pertaining to this lesion appear important. The review of the five cases previously described and the presentation of a new case, first to be diagnosed at the operating table, are the subject of this paper.

Review of Literature

Of the cases published, only five were found to fulfill the criteria of supravalvular aortic stenosis, and these five form the basis of this review.[2, 4, 6, 7, 11, 12] There were two males and three females in the group. All patients died between the ages of 30 and 48, with average survival of 39 years. In four cases, cardiac failure appeared to be the cause of death. One patient died of multiple emboli. The lesion was situated at the upper border of the sinus of Valsalva in all cases, and was described as a ridge in all but one, where it constituted a veritable membrane. Considerable left cardiac dilatation and hypertrophy was invariably present, with average weight of the heart being 580 Gm., and maximum weight reaching 1,250 Gm. Rheumatic endocarditis was found in one case, and associated cardiovascular anomalies in three (Marfans syndrome, bicuspid aortic valve, congenital aortic aneurysm). The aortic valve was grossly and microscopically normal in four cases. In one case, a bicuspid aortic valve, with thickening of the cups, was noted. In this case, the supra-aortic ridge was particularly well developed in the region above the missing cusp.

Clinical history was reported in three cases only, all of which presented congestive heart failure for variable periods of time. In one case only was chest pain a prominent feature. A systolic and diastolic murmur was noted over the precordium in two cases. It should be noted that in the case where no murmurs were heard, the patient was seen for the first time in shock and cardiac decompensation.

The microscopic appearance was discussed by two authors. Both describe the ridge as composed of connective tissue, with overgrowth of elastic fibers according

* Submitted for publication July 8, 1958.

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This work was supported in part by funds from U.S.P.H., H.T.S. 5206.

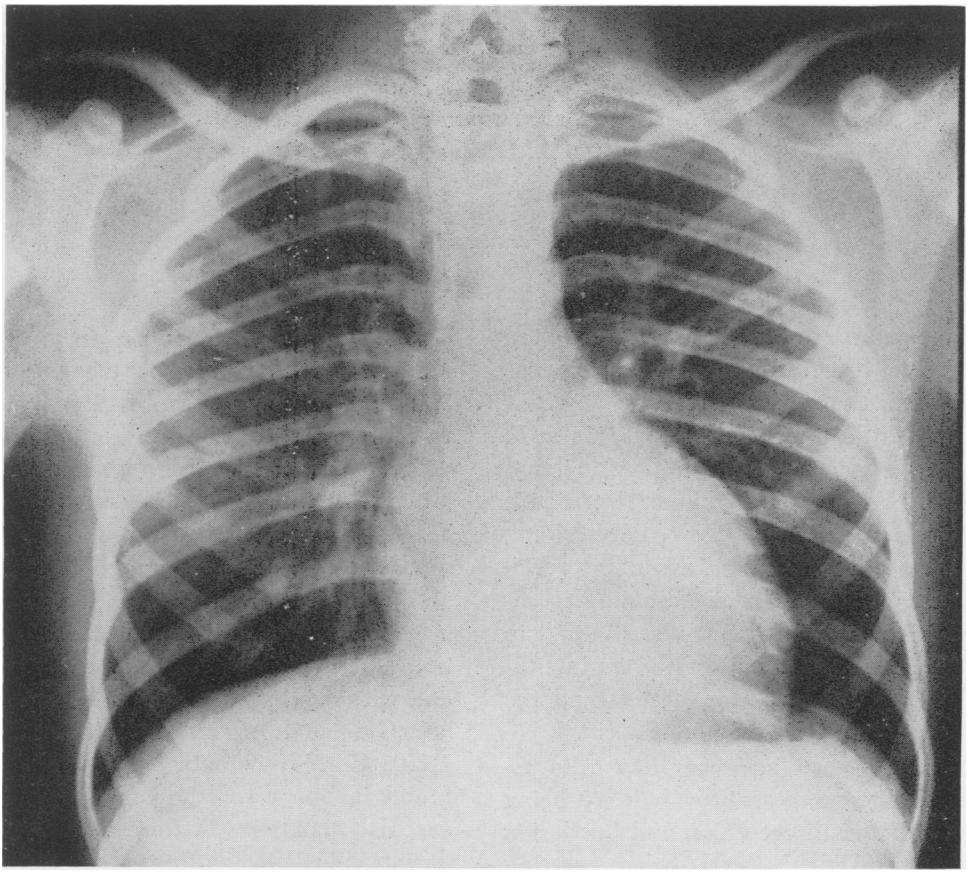

FIG. 1. P.A. chest plate showing left ventricular hypertrophy.

to Mencarelli [8] and decrease of elastic fibers according to Burry.[3]

Case Report

Clinical History: The patient, R. F., #46757, was a 12-year-old white boy admitted to the Mount Sinai Hospital with a chief complaint of chest pain of 5 months' duration. The patient had a known aortic systolic murmur from 3 months of age, with radiographic evidence of left ventricular enlargement from 3 years of age. At 10 years of age an aortic diastolic murmur, in addition to the systolic murmur, was first detected.

For 5 months prior to admission, the patient had episodes of sharp stabbing chest pain while at play, and episodes of squeezing retrosternal discomfort while at rest.

Physical examination showed normal development. The heart was enlarged to the left, with the point maximum impulse in the 5th intercostal space, at the anterior axillary line. A systolic thrill was easily palpated at the left 2nd and 3rd intercostal spaces. Auscultation revealed a harsh, grade three systolic murmur in the second intercostal space at the right sternal border, with radiation throughout the chest and into the neck. A grade II early diastolic murmur was also heard in the third and fourth intercostal spaces at the left sternal border. The pulse was 100 per minute, and the B.P. 95/45. Peripheral pulses were easily palpable. There was no evidence of congestive failure, cyanosis, or clubbing.

Laboratory studies of blood and urine were normal.

Chest x-rays showed enlargement of the left cardiac border suggestive of left ventricular hypertrophy (Fig. 1).

E.C.G. findings were those of left ventricular hypertrophy, i.e., a QRS of high voltage, an upright T wave in lead II and inverted T wave in leads III and AVF. The ST segment was depressed and the T wave diphasic in lead IV. These findings showed progression over the tracings of a year earlier.

A diagnosis of congenital aortic or subaortic stenosis was made.

Operative Findings: The patient was operated upon February 25, 1958, under general anesthesia. The chest was opened through a median marked sternotomy incision.

The left ventricle showed marked hypertrophy. The ascending aorta showed considerable post-stenotic dilatation, and a harsh systolic thrill could be palpated in its wall.

Following the introduction of catheters into the superior and inferior venae cavae and the femoral artery, and heparinization, the patient was placed on total cardiopulmonary bypass utilizing the De-Wall type bubble oxygenator and the Sigmamotor pump. Flow rates between 60 and 100 cc. per kilo. per minute were used to maintain an arterial mean pressure of 70 to 100 mm. Hg. Following the establishment of adequate perfusion, the aorta was cross-clamped and the heart arrested with acetylcholine (10 mg. per kg. body weight). The ascending aorta was opened by an incision into its anterior wall (Fig. 2).

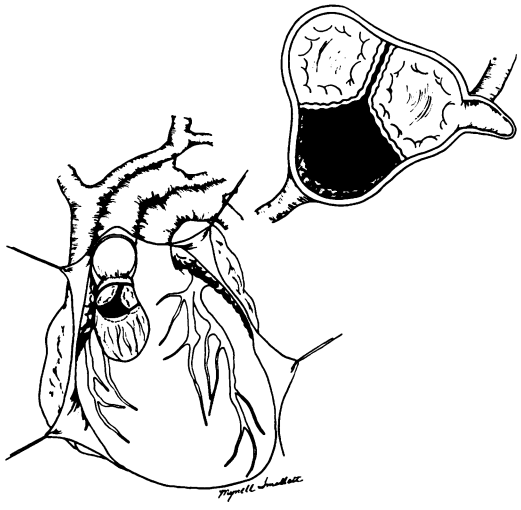

FIG. 2. The left ventrical and aortic outflow tract as seen at operation, showing the supravalvular aortic stenosis. Insert shows the deformity of the right coronary cusp.

Immediately distal to the cusps of the aortic valve, the aorta was found to have a slight annular constriction. This stenosis was reduced by repeated manual dilatation. Inspection of the aortic valve revealed the left coronary cusp, and the right non-coronary cusp to be present and of normal dimensions, although their borders appeared slightly furrowed and irregular. A normal right coronary cusp appeared absent, and the ostium of the right coronary artery could not be located. In the usual location of the right coronary cusp was found a hard fibrous elevation, firmly fixed to the side wall of the aorta. Direct manual palpation of the outflow tract of the left ventricle failed to reveal any sub-aortic stenosis. No further surgical procedure was undertaken. Air was evacuated from the left heart and aorta, and the aortotomy closed. Following this, the aortic cross-clamp was removed. The heart immediately picked up a spontaneous rhythm, and perfusion was discontinued. Total perfusion time was 60 minutes, with cardioplegia lasting 49 minutes. Shortly after perfusion the heart developed massive dilatation, followed by ventricular fibrillation. Effective electrical defibrillation was obtained, and a normal cardiac rhythm restored by manual massage. The heart again dilated and fibrillated. This was controlled by defibrillation and manual massage, but again recurred. In view of this inability of the heart to maintain effective action, catheters were again placed in the cavae, and the patient returned to the pump oxygenator. A careful search was made for any undiagnosed defects which might account for this picture, but none were found. It was noted at this time that if the aorta was cross clamped, the heart would not dilate, but that when the cross clamp was removed, the heart promptly dilated. Following a second perfusion of 41 minutes, efforts were again made to have the heart assume effective activity, but the heart developed intractible right and left sided dilatation, and failed to respond to intensive and prolonged massage. One hour after completion of the second perfusion, E.E.G. waves disappeared, and the patient expired.

Postmorten Examination: Description is limited to pertinent findings. The heart was markedly enlarged, weighing 360 Gm. (normal 125 Gm.). The enlargement was due to left ventricular hypertrophy and dilatation and slight dilatation of the left atrium. The site of the essential pathologic change was in the region of the aortic orifice. The aortic outflow tract was encroached upon by a bulging interventricular septum lined by smooth thin endocardium. The orifice was guarded by three semi-lunar cusps of approximately equal size. The cusps were irregularly thickened and fleshy and showed firm nodular excrescences projecting on both surfaces. On section, a gelatinous appearance was noted as well as dullness of the endocardial surface. The right coronary cusp was closely adherent to the aortic wall thus covering the orifice of the right coronary artery. The funnel shaped orifice of the left coronary artery was situated deep in the sinus of Valsalva. The aortic valve ring measured 5 cm. in circumference. Immediately above the sinus of Valsalva the aortic

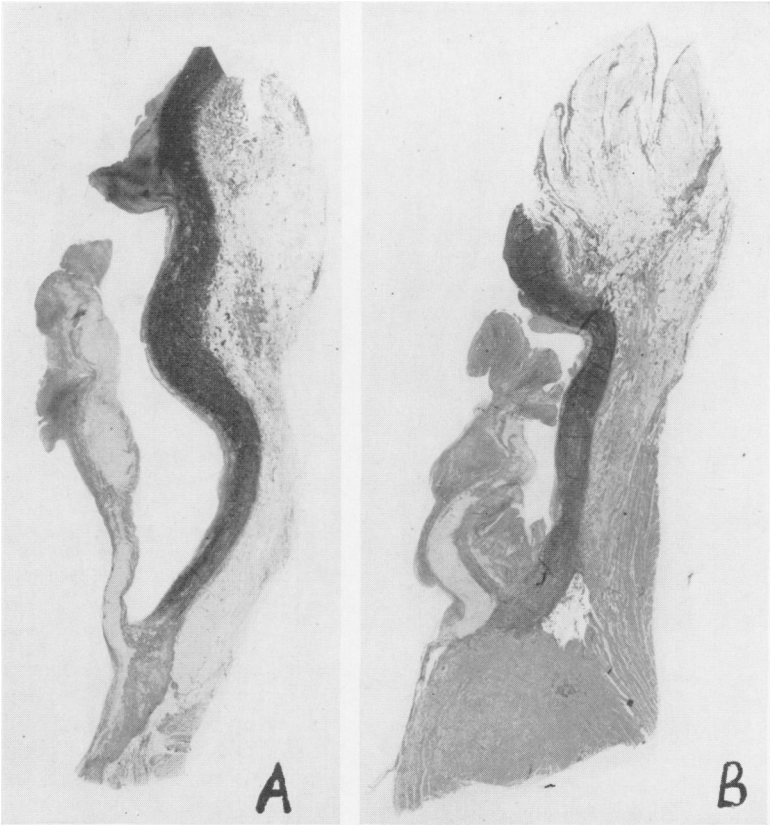

FIG. 3. A. The noncoronary cusp with overhanging supravalvular ridge. B. The right coronary cusp. Note thickening of the cusp and area of adhesion to the aortic wall. This adhesion has been forcibly torn during autopsy examination (Elastica-Van Geisen stain 1 : 1).

lumen was narrowed by a shelf-like circular ridge covered by intima. The width of the ridge was approximately 2 mm., and the circumference of the stenosing ring, 3.8 cm. Immediately above this, the circumference of the aorta widened to 4.8 cm.; there was no dilatation of the aorta past this point. A round orange-colored intimal plaque, about 1.5 cm. in diameter, was present in the ascending aorta just above the right sinus of Valsalva. The coronary arteries were patent, had normal distribution, and showed no evidence of atherosclerosis. Minimal streaks of fibrosis were noted on section of the left ventricular myocardium. The aorta was of normal size and showed minimal patchy intimal thickening in the thoracic portion.

Microscopic Findings: The myocardium, especially of the left ventricle, was hypertrophied. There were multiple small patchy areas of interstitial myocardial fibrosis, in both the anterior and posterior wall of the left ventricle. Occasional small perivascular scars were found in the myocardium. Minute, very recent foci of necrosis of individual fibers were present. The ventricular endocardium was thin, except for mild fibro-elastic thickening in the depth of the endocardial recesses in the left ventricle. There was fibro-elastic thickening of the left atrial endocardium.

Valve deformities were limited to the aortic valve. All three cusps were thickened due to increase in collagenous fibers and cellular connective tissue rich in basophilic ground substance. Intermingled with the collagenous fibers were scant, delicate elastic fibrils. The left coronary cusp was the least involved, the right coronary cusp the most severely thickened (Fig. 3 A, B). The valve deformity was accentuated by the presence of hyalinized collagenous verrucous excrescences, especially on the ventricular surface and on the edge of the cusp. In the right coronary cusp these were seen also on the inner aspect of the valve facing the sinus of Valsalva. While most of the nodules were avascular, one appeared split by a wedge-

shaped, vascularized band of connective tissue which faced the aortic wall without being attached to it at the time of examination. The corresponding intima of this sinus of Valsalva was likewise occupied by nodular fibrous excrescences which in the available sections contained no blood vessels. In the other valve pockets the aortic intima was not thickened at that level. A sagittal section through the stenosing ring at the upper border of the sinus of Valsalva showed that the projecting structure was continuous with the aortic media. On section it appeared like a hillock composed predominantly of collagenous tissue, with elastic fibers fanning out from the media and traversing it in various directions. Occasional tiny vessels were present. The ridge was confined within the limits of the internal elastic lamella. The intima over both slopes of the ridge was thickened. The intimal plaque in the ascending aorta consisted of fibroelastic tissue and foam cells. Except for scattered small intimal atheromatous patches in the thoracic aorta there were no other significant alterations of the aorta. The coronary arteries were normal.

Discussion

The stenosing ring, constituting the essential lesion in this case, appears to be congenital in origin. Study of the same area in normal hearts reveals a small invagination of the aortic wall normally present at the upper limit of the sinus of Valsalva (Fig. 4). It can be postulated that the supra-aortic valvular stenosis is a developmental exaggeration of this normal structure. The architecture of this lesion and the outgrowth of the stenosing ring out of the media bear a certain resemblance to the microscopic structure of the infantile coarctation of the aorta.[5]

The thickening of the cusps and the adhesion of one of them to the aortic wall raises the possibility of a superimposed inflammatory episode, which would by the same token explain the acute onset of symptoms relatively early in life. The microscopic examination, however, disclosed nothing to confirm this idea. Whether the changes observed in the cusps could have resulted from the mechanical insult created by the abnormal hemodynamic conditions, is subject to debate.

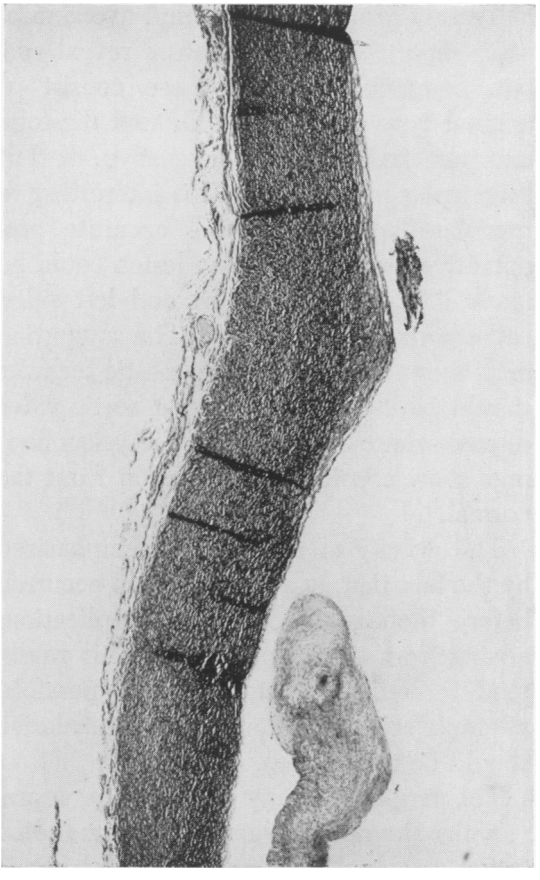

FIG. 4. Supravalvular region in a normal baby. Note invagination of the aortic wall at the upper border of the sinus of Valsalva (Weigert-elastica stain 1 : 20).

Another possible explanation would be to ascribe the definite thickening and deformity of the aortic cusps to an inborn derangement of the mesenchymal tissue to be considered as a separate and concomittant congenital anomaly. Similar interpretation was given by Prior [9] to valvular lesions associated with endocardial fibro-elastosis. The operative dilatation of the stenosing ring performed in this case produced prompt cardiac dilatation on removal of the aortic cross clamp. This can be explained by the massive aortic insufficiency resulting from the fixation of the right aortic cusp to the aortic wall in the open position. It is felt that in this case the stenosing ring limited the degree of insufficiency and its removal resulted in overwhelming regurgitation.

A study of our own case and five similar cases reported in the literature reveal certain common features. These consist of anginal type of pain, systolic and diastolic murmur and findings suggestive of left ventricular hypertrophy. It is interesting to speculate whether a more accurate preoperative assessment of the lesion could be made if angiocardiography and left sided catheterization were used. The suggestion may be made that these diagnostic technics should be used in any case of aortic valve disease where the history and physical findings show considerable variation from the normal.

The gravity of the lesion is emphasized by the fact that, in all cases, death occurred before the age of 50 from complications arising from valvular anomaly. This would tend to indicate that, whenever possible, attempts at surgical correction of this lesion should be undertaken.

The surgical therapy of choice for supravalvular stenosis appears to be manual dilatation or actual excision of the obstructing lesion if the latter appears possible.

Summary

Five cases of supravalvular aortic stenosis were collected from the literature and reviewed. A new case of supravalvular stenosis, in which an attempt at surgical relief of the condition was made, is presented. The nature and the surgical implications of this lesion are discussed.

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