Acquired Pulmonic Stenosis
Report of a Case Caused by Mediastinal Neoplasm

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Although congenital pulmonic stenosis is a common cardiac lesion, acquired pulmonic stenosis is rare. We have recently had the opportunity to study a case in which the physical findings strongly suggested pulmonary stenosis. It was ultimately determined that the obstruction of the right ventricular outflow tract was caused by lymphoblastoma of the anterior mediastinal lymph nodes.

Case Report
A 15-year-old girl was referred to the University of Michigan Medical Center for evaluation of a systolic murmur that had been discovered by her local physician during a routine physical examination. Multiple previous examinations had disclosed no evidence of heart disease. There was no history suggesting rheumatic fever, and the patient had never received radiation therapy. A system review was negative except for a five-pound weight loss over the previous two months.

The patient was a healthy, well-nourished young girl. The blood pressure was 110/80 mm. Hg. All peripheral pulses were strong and equal. There was no venous distention, and the lungs were clear. A localized systolic lift was detected over the second left interspace, but there were no palpable thrills. The heart was not enlarged to percussion. On auscultation a grade 4/6 harsh, systolic ejection murmur was noted in the pulmonic area. The murmur was widely transmitted over the left upper chest and could be heard posteriorly over the seapula. The pulmonic component of the second sound was diminished in intensity so that at times only a single component could be heard in the pulmonic area. At other times physiologic splitting was identified. The other findings in the cardiac examination were within normal limits. Neither the liver nor the spleen was palpable on the initial examination.

The hemoglobin level was 13.6 Gm. per cent and the leukocyte count was 9,200. Results of urinalysis were normal. Frontal and lateral films of the chest showed an anterior mediastinal density which projected primarily to the left of the cardiac silhouette (fig. 1).

An electrocardiogram showed right axis deviation but was otherwise within normal limits. A phonocardiogram demonstrated the systolic ejection murmur (fig. 1). Cardiac catheterization showed no evidence of shunt. All oxygen saturation determinations were within normal limits. The right ventricular systolic pressure was between 40 and 50 mm, and the pulmonary arterial pressure was about 15 to 20 mm. A short infundibular chamber was traversed by the catheter (fig. 2). Angioarthrography with injection of 40 ml. of Ditriol onto the right ventricle demonstrated compression of the base of the heart by the mediastinal mass with resultant infundibular pulmonic stenosis and posterior displacement of the pulmonary artery and aorta. It also demonstrated the nonvascular nature of the mass.

Shortly after admission the spleen tip was palpated. During the ensuing week the spleen enlarged rapidly to 3 cm. below the costal margin. Although there were no palpable nodes, a routine supravalvular biopsy disclosed a small scalene node with large immature lymphocytes replacing the normal architecture. The diagnosis of lymphoma of the lymphocytic type was made.

The patient received four intravenous doses of nitrogen mustard over an 8-day period. Following therapy there was a marked reduction in the size of the mediastinal mass radiographically, and the systolic murmur disappeared completely (fig. 1).

Discussion
Although a mediastinal neoplasm was immediately suspected in this case, the possibility of pulmonic stenosis with an unusual degree of poststenotic dilatation of the pulmonic artery was considered as a remote possibility. After the nonvascular nature of the mass had been established, the most likely possibility was lymphoma, although benign teratoma and thymoma were hopefully considered. The diagnosis was confirmed by bi-
ops and by the dramatic response to nitrogen-mustard therapy.

The degree of outflow obstruction in this case raises the question of pericardial invasion. After a brief remission this patient showed radiographic evidence of massive regrowth of the mediastinal tumor without again developing a systolic murmur. We have seen other cases in which neoplasms have greatly compressed and displaced the heart without producing signs of obstruction of the great arteries.

Mediastinal neoplasms frequently obstruct the superior vena cava; they rarely obstruct the great arteries. To our knowledge, only five cases of pulmonary stenosis produced by extrinsic compression have been reported.1-5 One such case reported by Winters1 was due to the involvement of the anterior mediastinal lymph nodes by Hodgkin's disease. Compression of the pulmonary artery was demonstrated angiographically. Waldhausen and co-workers2 reported the typical murmur of pulmonic stenosis produced by compression of the right ventricle and pulmonary artery by a primary mesothelioma of the pericardium. The murmur disappeared following resection of the tumor. Three cases of teratoma have been reported3-5 in which the growth appeared in the anterior mediastinum and produced pulmonic stenosis by compression of the pulmonary artery. In two cases

Circulation, Volume XXVI, November 1962

Figure 1

Phonocardiogram and posteroanterior film of chest before and after nitrogen mustard therapy. Note the decrease in the size of the mediastinal silhouette and the disappearance of the systolic murmur on the phonocardiogram.
ACQUIRED PULMONIC STENOSIS

Figure 2

Lateral x-ray of chest during angiocardiography, demonstrating the smooth contoured infundibular narrowing. At cardiac catheterization the pressure gradient across the infundibular chamber was demonstrable. The pressure in the right ventricular cavity was about 40 mm.; distal to the obstruction it dropped to 15 mm.

resection of the lesion resulted in the disappearance of the cardiac abnormalities.

Autopsy evidence of pulmonary stenosis has been reported in cases of far-advanced pulmonary tuberculosis. All such cases (four in number) were associated with a generalized miliary process; correlation of autopsy findings with previous clinical observations was not reported, other than mention of a soft systolic murmur over the base of the heart. Rheumatic pulmonary valve stenosis is extremely rare and is invariably associated with mitral and tricuspid disease. Cases have been reported by Herbst, McGuire, and others. In 1937 Gouley reported five cases of rheumatic pericarditis causing supravalvular stenosis. The narrowing occurred 2 cm. above the pulmonic valve at the reflection of the pericardium, and in each case the stenosis was associated with a harsh systolic murmur, a thrill, and an increase in the pulmonic second sound. The diagnosis in these cases was confirmed at autopsy, which showed definite evidence of the rheumatic origin. Bacterial endocarditis affecting previously normal pulmonary valves has been reported to cause stenosis in four autopsy-confirmed cases, all of which occurred before the advent of antibiotic agents. Degenerative disease is even more rare as a cause of pulmonary stenosis. Doniger described a single case of atherosclerotic disease of the pulmonary artery involving the valve cusps, and Bufalini reported an aneurysm of the ascending aorta that compressed the pulmonary artery and gave clinical findings of pulmonic stenosis. Primary tumors have occasionally been reported to involve the pulmonary valve. In 1955 Richmond described a case of myxoma of the pulmonary valve and cited five other cases in the literature, all of which were based primarily on autopsy findings rather than the clinical picture.

One of the most interesting varieties of acquired pulmonic stenosis is that associated with malignant carcinoid tumors. In 1943 Millman reported a case associated with liver metastases. Fibrosis of the endothelium of the right heart was demonstrated with fusion of the pulmonic valves, which otherwise were normal. A review of several similar cases has been published. Most of these have shown involvement of both the pulmonary and the tricuspid valves. Pathogenesis of these lesions

Circulation, Volume XXVI, November 1962
is probably related to the vasoconstrictive properties of serotonin secreted by the carcinoid tumor.

Summary

Acquired pulmonic stenosis is rare. A well-documented case of pulmonary infundibular stenosis due to mediastinal lymphoma has been described. Complete disappearance of the heart murmur followed therapy with nitrogen mustard. Other causes of acquired pulmonic stenosis have been discussed.

References


Religio Medici

I could never divide my self from any man upon the difference of an opinion, or he angry with his judgment for not agreeing with me in that from which perhaps within a few days I should dissent my self.—Sir Thomas Browne. Religio Medici. Edited by W. A. Greenhill, M.D. London, MacMillian and Co., Ltd., 1950, p. 12.

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