CLINICAL PROBLEMS IN CARDIOPULMONARY DISEASE

Acquired Pulmonic Stenosis due to a Mediastinal Seminoma *

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A 43-year-old man was referred for the evaluation of an apparently new cardiac murmur and a mediastinal mass on the chest x-ray film. Five weeks before admission, he had noted the insidious onset of a persistent nonproductive cough, intermittent fever to 38.5°C (101.3°F), and dull, aching, nonproductive nonpleuritic discomfort in the chest. The patient had consulted his private physician, who had noted a cardiac murmur which had not been present during a routine physical examination two years previously. A chest x-ray film demonstrated a mediastinal mass, and the patient was referred for further evaluation.

Examination disclosed a healthy appearing man in no distress. Pertinent physical findings included a prominent “a” wave in the jugular venous pulse. There was a grade 3/6 long crescendo-decrescendo murmur that was best heard at the pulmonic area and upper left sternal border. The murmur varied little, if any, with respiration, and there was no change with Valsalva’s maneuver. The second heart sound was thought to be single, without inspiratory splitting. There was no ejection click. The chest was clear to auscultation and percussion, without localized wheezing. There was no hepatosplenomegaly, abdominal or testicular masses, or palpable lymphadenopathy.

The chest x-ray film obtained on admission demonstrated a 10 cm × 8 cm lobulated mass in the anterior mediastinum. (Fig 1). The electrocardiogram was normal. The results of routine laboratory studies were normal. Fluoroscopic examination showed the mass to be nonpulsating and distinct from the intrathoracic vasculature. A computerized tomographic scan of the thorax demonstrated in the region of the pulmonary outflow tract a soft tissue mass contiguous with the pericardium and extending into the left hilum. A phonocardiogram obtained on admission (Fig 2) confirmed the initial auscultatory findings. An echocardiogram demonstrated a normal right ventricular dimension and septal motion. The pulmonic valve could not be visualized.

A thoracotomy was performed. The large mediastinal mass was found to be invading the main pulmonary artery, left hilum, and pericardium. There were numerous implants of tumor on the epicardial surface. The mass was partially resected and pathologically was a pure seminoma.

Once the diagnosis of a mediastinal seminoma was made, a liver-spleen scan, bone scan, and lymphangiogram were performed. All showed normal findings. The concentrations of a-fetoprotein and ß-human chorionic gonadotropin were not elevated. The patient underwent a full course of irradiation,

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Figure 1. Posteroanterior (top) and lateral (bottom) chest x-ray films taken on admission reveal large anterior mediastinal mass.
Discussion

Acquired pulmonic stenosis is relatively unusual.\textsuperscript{1} Reported causes include metastatic carcinoid syndrome, rheumatic heart disease (in which involvement of the other cardiac valves is nearly always present), bacterial endocarditis in either congenitally deformed valves or de novo in narcotic addicts, rare primary tumors of the valve (including sarcoma and myxoma), and extrinsic lesions (including anterior mediastinal tumors, ascending aortic aneurysms, aneurysms of the sinus of Valsalva, and constrictive pericarditis).

In this case, pulmonic stenosis was produced by an anterior mediastinal seminoma compressing the main pulmonary artery and invading the pericardium. Anterior mediastinal tumors causing pulmonic stenosis are rare. There are approximately 25 cases reported in the literature.\textsuperscript{2,4} Most tumors to date have been either teratomas or Hodgkin's disease. Other reported tumors include thymoma, lymphatic cyst, pericardial mesothelioma, intrapericardial sarcoma, and lymphoblastoma. This appears to be the first reported case of a seminoma causing pulmonic stenosis.

Most of the patients initially had symptoms of pain in the chest, dyspnea on exertion, and cough, although occasionally asymptomatic patients were discovered by accident. There are several auscultatory characteristics discernible from the reported cases of pulmonic stenosis secondary to mediastinal tumors. The murmur of pulmonic stenosis is generally of grade 2-3/6 in intensity. Characteristically, there is no associated systolic click. Littler et al\textsuperscript{2} emphasized the respiratory variation of the murmur. In three of four of their patients, the murmur markedly diminished on inspiration and was of

\textbf{FIGURE 2.} Phonocardiogram obtained on admission demonstrates long crescendo-decrescendo systolic murmur (SM) recorded both at pulmonic area (PA) and at apex. Note absence of ejection click and appearance of late, soft pulmonic second sound (P\textsubscript{2}) at pulmonic area. 1, first heart sound; A\textsubscript{2}, aortic second sound; and MF, medium frequency. Adding 4,025 rads to the mediastinum, with disappearance of the mass. At a follow-up examination after completion of irradiation, no murmur was detectable. The second heart sound was now physiologically split. The murmur's disappearance was demonstrated by the phonocardiogram (Fig 3).

\textbf{FIGURE 3.} Phonocardiogram after treatment of seminoma. Murmur of pulmonic stenosis has disappeared. PA, Pulmonic area; and MF, medium frequency.

494 KINDIG, TAVEL

CHEST, 78: 3, SEPTEMBER, 1980
maximal intensity during full expiration. These investigators postulated that this variation was due to the lifting of the tumor off the pulmonary artery during inspiration and increased compression during expiration. The second heart sound is variably reported to be normally split or single and of increased, normal, or decreased intensity. A prominent "a" wave in the jugular venous pulse is occasionally mentioned. Findings of right ventricular pressure overload and overt right ventricular failure appear to be related to the duration and severity of the compression of the pulmonary outflow tract.

Most of the patients with this condition have undergone right heart catheterization and pulmonary angiographic studies. These procedures have demonstrated extrinsic compression of the main pulmonary artery and a systolic pressure gradient of 10 to 100 mm Hg between the right ventricle and pulmonary outflow tract. We believe that the use of these procedures in the case reported herein was not necessary. An anterior mediastinal mass was demonstrated in the area of the pulmonary outflow tract by chest x-ray film and computerized tomographic scanning of the thorax. A thoracotomy was necessary for histologic diagnosis. With primary treatment of the tumor, the murmur of pulmonic stenosis resolved.

Although relatively unusual, an adult patient with auscultatory findings characteristic of pulmonic stenosis, especially with no known previous cardiac disease or cardiac murmur, should be evaluated carefully for the various causes of acquired pulmonic stenosis listed previously. If there is no systolic click or regurgitant murmur and if the stenotic murmur increases with expiration and diminishes with inspiration, an associated anterior mediastinal mass would be the most likely diagnostic possibility.

REFERENCES