9-1-1986

Left Atrial Myxoma with Coexistent Lung Carcinoma: Report of a Case and Review of the Literature

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Left atrial myxoma with coexistent lung carcinoma: Report of a case and review of the literature

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A case of left atrial myxoma in a patient with a long history of smoking who also had carcinoma of the lung is reported. Both diseases were diagnosed at autopsy. The sometimes subtle clinical presentation of atrial myxoma, the diagnostic techniques, its management and its histogenesis are discussed. The possibility of smoking as a risk factor is speculated upon.

Primary neoplasms of the heart are rare, with an incidence of up to 0.05 percent at autopsy.[1-3] Atrial myxomas, the most common primary heart tumors, are left sided in 75 percent of cases; the incidences of right atrial and ventricular myxomas are 20 percent and 5 percent, respectively.[1,4] The risk factors are not known. The clinical features of the tumor vary considerably from case to case—some tumors have been asymptomatic, others have produced severe valvular obstruction, and yet others have, without prior warning, caused sudden death. Although atrial myxoma can be demonstrated by both selective angiocardiography and echocardiography, on occasion it is an unexpected finding during mitral commissurotomy or at autopsy. This tumor also poses some problems pathologically. Despite several histochemical and ultrastructural studies of cardiac myxomas, the progenitor cell of this lesion remains controversial. Many authors believe it to be a true neoplasm that originates from an endocardial cell, while others maintain that the tumor is an organizing thrombus.[1]

The following case is being reported for several reasons: first, to stress the subtle clinical presentation of atrial myxomas; second, to discuss management; third, to discuss histogenesis, with particular attention given to the presence of factor VIII antigen; and, fourth, to point out the coexistent lung carcinoma. We were unable to find any previous report of coexisting cardiac myxoma and adenosquamous cancer of the lung in the English-language literature.

Report of case

An 80-year-old Caucasian man arrived at the emergency room with chief complaints of chest pain, weakness, syncope, and tarry stools, all of which had been present for 3 days. The patient's medical history included myocardial infarction 10 years earlier, hypertension, chronic obstructive pulmonary disease, and peptic ulcer disease, which had in the past necessitated transfusion. He reported a 3-year history of dyspnea on exertion when walking across the living room. He had a 60-pack year smoking history and was an ethanol abuser. The patient stated that he had lost 30 pounds in the 6 months prior to admission.

Physical examination on admission revealed an alert, oriented, emaciated elderly man who was in respiratory distress. The trachea deviated to the right, and the right lung was dull to percussion. Increased anteroposterior diameter of the chest and pectus excavatum deformity was noted. Breath sounds were markedly decreased bilaterally, with total absence of breath sounds at the right lung base. Wheezes were heard in the right upper lobe. The cardiac rhythm was irregular at a rate of 120 beats per minute, with muffled heart tones. Bilateral pedal edema was observed.

Laboratory studies revealed the following values: hemoglobin concentration, 6.1 gm./dl.; hematocrit, 19.8 percent; blood urea nitrogen, 76 mg./dl.; serum creatinine, 2.6 mg./dl.; and serum albumin, 2.4 gm./dl.

The electrocardiogram revealed atrial fibrillation with rapid ventricular response, myocardial ischemia, left axis deviation, left anterior hemiblock, and low voltage of QRS complexes. Chest roentgenography demonstrated generalized cardiomegaly, pulmonary vascular congestion, and a 5-cm. irregular infiltrate in the right upper lobe, with coexistent right pleural effusion.

With the patient hypotensive secondary to the gastrointestinal hemorrhaging that was in process, initial emergent management involved placement of large-bore peripheral and central intravenous lines. Supplemental oxygen was provided for hypoxia, and crystalloid infu-

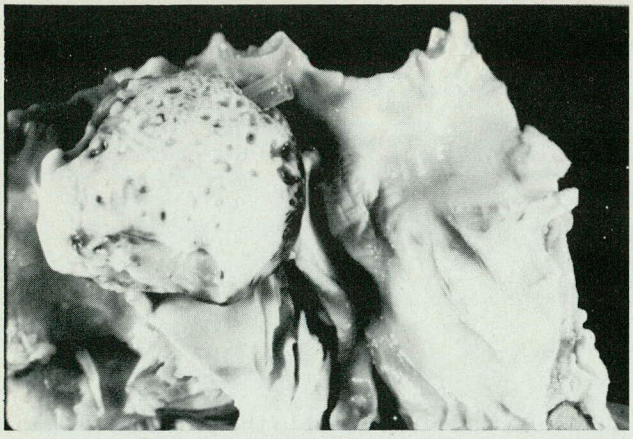


Fig. 1. *A soft, pedunculated, gelatinous, partly smooth-surfaced, yellowish-tan mass measuring 5 × 5 × 5 cm. was found attached to the left atrial lateral wall at the time of autopsy.*

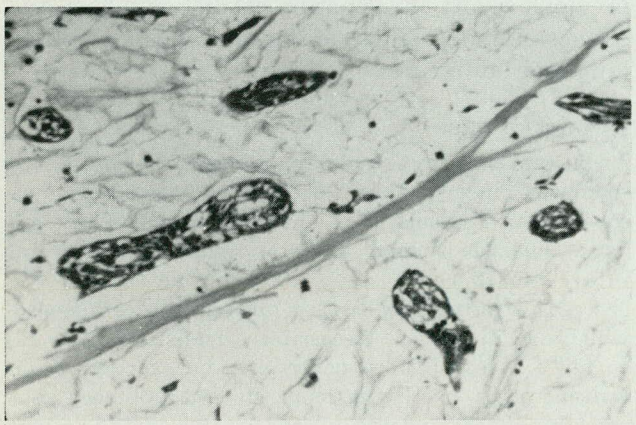


Fig. 2. *Groups of polyhedral to round cells forming duct-like spaces and cords intermixed with vascular structures embedded in a myxoid stroma were seen.*

sion was begun at a rapid rate, which corrected the hypotension. A nasogastric tube was placed and iced saline lavage was performed, but this procedure failed to slow the active gastrointestinal bleeding. Transfusion was begun as soon as blood products were available.

The patient was transferred to the intensive care unit. Because of the extreme seriousness of his condition, immediate surgical consultation was sought and arrangements were made for immediate operative intervention. Once in the surgical suite, prior to the initial incision, the patient experienced cardiac arrest. He was revived, was returned to the intensive care unit, suffered another arrest, and, despite extensive resuscitative attempts, died. Permission for autopsy was requested and granted.

Autopsy findings

Gross inspection. Thoracic dissection revealed right pleural adhesions, with minimal bilateral pleural effusion. Both lungs weighed 540 gm. An intrapulmonary neoplasm in the right upper lobe measured 5.0 cm. in diameter. On its cut surface, the mass was tan/grey and soft. The rest of the lung tissue was anthracotic, with multiple areas of emphysematous changes and interstitial fibrosis.

The heart weighed 300 gm., and there was 500 cc. of clear pericardial fluid. The predominant right coronary arterial system showed focal yellow calcific atheroma producing insignificant stenosis of the lumen. The endocardial surfaces and leaflets of valves were smooth except for the left atrium, which contained a 5 × 5 × 5 cm. pedunculated mass firmly attached to the left atrial wall (Fig. 1). The mass was soft, gelatinous, partly smooth, and yellow to tan in color, with areas of hemorrhage and calcification. Valve diameter measurements were as follows: tricuspid, 10.0 cm.; pulmonic, 7.0 cm.; mitral 8.0 cm.; and aortic, 6.5 cm. The left ventricle measured 2.0 cm. and the right ventricle 0.3 cm. in thickness.

There were benign duodenal and gastric ulcers, superficial erosions of the stomach, multiple benign colonic polyps, and multiple sites of metastatic carcinoma in the liver. The genitourinary system showed arterionephrosclerosis, chronic pyelonephritis, prostatic hypertrophy, and cystitis cystica.

The brain weighed 1,130 gm. and exhibited atrophy but no infarction. No thrombi were seen in any of the organs.

Light microscopic examination. Microscopic study of the lung tumor and its metastatic foci in the liver revealed a moderately differentiated adenosquamous carcinoma.

Examination of the atrial mass showed a myxoid neoplasm with variable histology. There were areas where single-layered cells, the shape of which varied from round to polyhedral to stellate, formed cords (Fig. 2). There were other areas of branching and nonbranching vessels composed of several layers of spindly tumor cells surrounding well-defined spaces filled with erythrocytes and leukocytes. The stroma of the tumor was myxoid in some areas and fibrous in others. These findings were judged to be compatible with a diagnosis of atrial myxoma.

Discussion

Diagnosis

Signs and symptoms. The clinical presentation of atrial myxoma may be classified into three separate types, as described by Peters and associates[5] and Paulsen and coworkers[6]: (1) mechanical effects caused by obstruction of blood flow; (2) constitutional abnormalities similar to those seen in most systemic diseases; and (3) symptoms resulting from emboli released into the pulmonary or systemic arterial beds.

Symptoms of mitral stenosis-like disease include fatigue, dyspnea on exertion, and chest pain, while sudden occlusion of the mitral orifice with a ball-valve blockade produces acute paroxysmal dyspnea, cough, acute pulmonary edema, hemoptysis, and syncope. Nearly 90 percent of patients in these authors' studies presented with systemic illness, which was characterized by fever, weight loss, anemia, fatigue, elevated erythrocyte sedimentation rate, and elevated immunoglobulins values. Forty

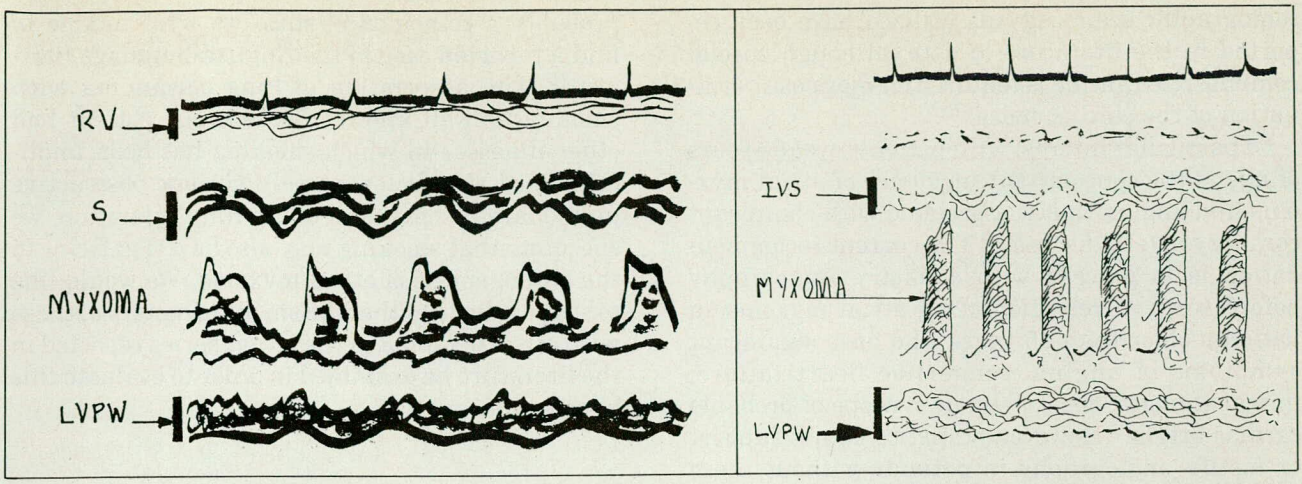
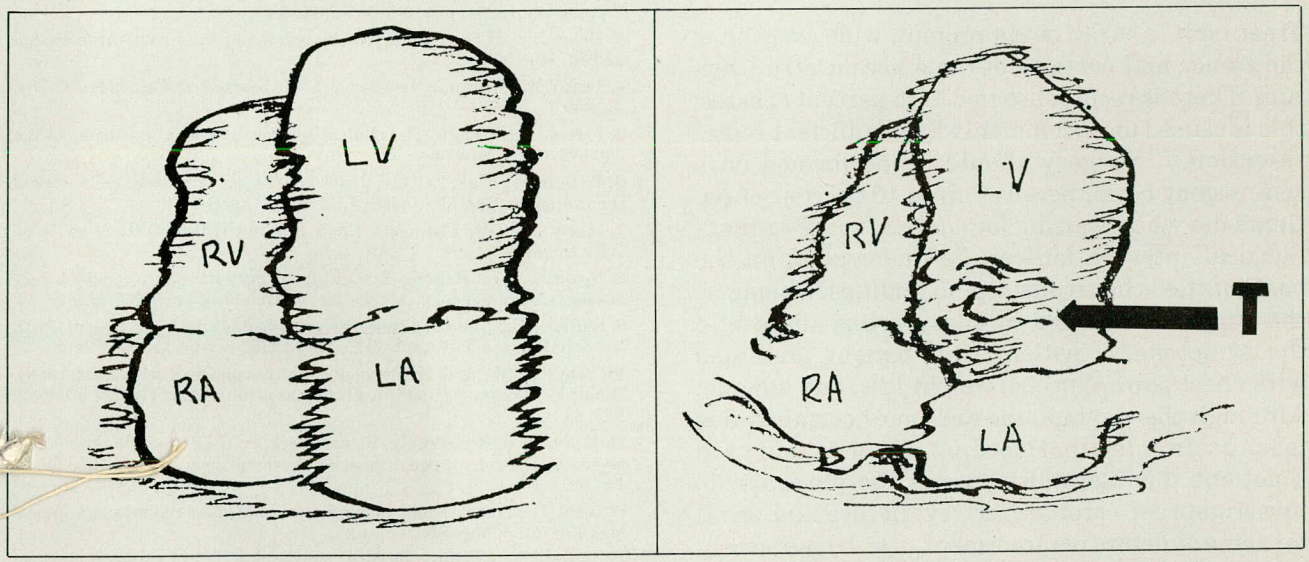


Fig. 3A. *Prominent left atrial myxoma as noted on standard M-mode echocardiogram (RV = right ventricle, S = interventricular septum, and LVPW = left ventricular posterior wall).* Fig. 3B. *Prolapsing left atrial myxoma as seen in diastole from an M-mode echocardiogram (IVS = interventricular septum).*



Figs. 4A and 4B. *Parasternal long-axis view 2-D echocardiogram showing normal anatomy (A) and left atrial myxoma (B) (T = tumor, LV and RV = left and right ventricles, and LA and RA = left and right atria).*

to 50 percent of patients present with signs of embolization, most frequently to the brain, kidney, and extremities. Frequently dizziness and syncope are reported, and sudden death has occurred.[7]

Signs of this tumor vary from an accentuated, split S1 sound to an accentuated P2 of the second heart sound. Apical systolic or diastolic murmurs, or both, have also been described. While positional variation of symptomatology is helpful in the diagnosis of atrial myxoma, it is a relatively rare physical finding.[2,6-8] Electrocardiologic characteristics are minimal and include sinus rhythm, left atrial enlargement, left ventricular hypertrophy, and, rarely, atrial fibrillation.

Radiologic procedures. Salcedo and coworkers[9] presented 25 cases of atrial myxoma, only 3 of which were diagnosed before the use of echocardiography. Standard M-mode echocardiography showed the characteristic mass and decreasing E-F slope (Figs. 3A and 3B). Lesions missed by this technique were found on 2-dimensional echocardiography, which is the procedure of choice for diagnosis of this mass (Figs. 4A and 4B).[2,6,7,9,10] Transesophageal echocardiography has demonstrated cardiac lesions in patients who had poor conventional results.[11] This technique may become more important in diagnosing a myxoma in the older patient with a limited "echo-window" and increased lung shadowing because of chronic obstructive pulmonary disease.

Three cases of atrial myxoma in which computed

tomographic scanning was utilized have been reported in the literature to date, although special computerized gating is required for diagnostic evaluation of the cardiac mass.[12,13]

Of particular interest with patients over 50 years of age is the concomitant incidence of atrial myxoma and angina that is associated with significant coronary artery disease.[8,9] The current recommendation is to proceed with coronary angiography before surgical resection of an atrial myxoma in patients older than 35 years who have significant symptoms of angina, congestive heart failure, mitral stenosis, chest pain, or syncope of probable cardiac origin.[8] However, statistics do not support or justify angiography in patients without these symptoms.

Treatment

Treatment is curative via prompt, wide excision of the tumor and portions of the adjacent atrial septum. There is recurrence in 5 to 15 percent of cases; this is caused most commonly by insufficient septal resection.[1,7] Surgery should be performed on a semiurgent basis, because up to 10 percent of patients die while waiting for surgical intervention.[7] Surgical intervention can be undertaken on the basis of the echocardiographic findings, so long as the surgeon considers coronary artery disease in the symptomatic patient. Our patient presented with chest pain, syncope, weight loss, and anemia. Although these symptoms were most certainly due to his gastrointestinal bleeding, historic findings in a patient this age must prompt the physician to investigate for coronary artery disease and atrial myxoma if definitive treatment is to be pursued.

Histogenesis

Despite several histochemical and ultrastructural studies of cardiac myxomas, the cell of origin and certain structural features of this tumor remain subjects of controversy. Many doubt a neoplastic nature and prefer a thrombotic origin.[14] However, according to the histogenetic assessment by Morales and associates,[15] the endothelial cell is the most likely progenitor cell. These authors believe that this tumor is indeed a neoplasm. Factor VIII, which is present in the majority of these tumors, further confirms their endocardial origin.

Conclusions

The coexistence of atrial myxoma and adenosquamous carcinoma of the lung in this patient is probably a coincidence, since we were unable to find a reported case in the English-language literature. The association of lung carcinoma with smoking is well known. Because our patient had other illnesses in which smoking has been implicated as a risk factor, namely, chronic obstructive pulmonary disease, and myocardial infarction, we speculate that smoking may also be a risk factor in the pathogenesis of atrial myxoma. We would like to suggest further that the smoking history of those with atrial myxoma in the larger series reported in the literature be examined in order to evaluate this hypothesis.

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Accepted for publication in March 1986. Updating, as necessary, has been done by the authors.

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