pulmonary artery pressure 70/28 mm Hg, with a mean of 42
mm Hg, pulmonary capillary wedge pressure 24 mm Hg, left
ventricular pressure 155/22 mm Hg, aortic pressure 155/80
mm Hg, and aortic mean pressure 108 mm Hg. The cardiac
index was 3.04 l/m². The end-diastolic volume was in-
creased. The ejection fraction was 0.34.

A left ventricular angiogram revealed a dilated left ven-
tricular cavity with generalized severe hypokinesia. The left
ventricular wall thickness was of normal size.

Coronary angiography revealed no abnormalities of the
coronary arteries with delayed clearance of dye (Fig 3 and
Fig 4).

DISCUSSION

In 1943, Weiss⁵ based his description of scleroderma
heart disease on two necropsy patients who demon-
strated atypical patchy myocardial fibrosis, normal
extramural coronary arteries and intimal proliferation of
the small coronary vessels. Subsequently, these findings
have been confirmed by other reports.¹-³

Since scleroderma patients have diffuse alteration of
myocardial small vessel disease, functional derangements
in myocardial microcirculation and possible impairment
of capillary transport may explain their heart disease.

A similar view was expressed by Tambe et al⁴ in
explaining delayed clearance of dye observed angiog-
raphically in four patients with angina pectoris, normal
coronary arteries and myocardial involvement (dilatation
and hypokinesia in two patients). Increased resistance of
the capillary bed, regardless of etiology (arterioscle-
rotic heart disease, diabetes mellitus, certain cardio-
myopathies, etc) and in our patient, scleroderma could
lead to myocardial dysfunction.

We presented evidence⁵-⁷ that diabetics can develop
myocardial disease without large coronary artery in-
volvement (diabetic cardiomyopathy), possibly due to
pathologic changes in small coronary vessels.

Our patient presented myocardial dysfunction docu-
mented by noninvasive as well as by direct methods.

The weak first heart sound, the presence of S₂, gallop,
the abnormal systolic time intervals point in this direc-
tion. The vectorcardiogram showing infero-anterior
myocardial damage, while the coronary arteries were
normal, suggests the possibility of diffuse impairment of
the microcirculation. Hemodynamic studies revealed bi-
ventricular dysfunction and left ventricular hypokinesia.

In the majority of patients who develop congestive
heart failure, most cases are due to cor pulmonale, sys-
temic hypertension secondary to "scleroderma kidney" or
pericardial disease⁸ with intrinsic myocardial disease
making up a very small number. Although cardiomegaly
is demonstrated in these patients, only rarely, is it ever
massive.²

Histologic examination usually revealed only mild
noninflammatory intimal thickening of coronary vessels
supporting the hypothesis that fibrosis results from heal-
ing of microscopic infarcts secondary to the narrowing of
these small arteries and arterioles. Occlusion of the major
coronary vessels is rare in scleroderma and typical myo-
cardial infarction was reported in only 9 of 275
patients.²

It is tempting to speculate that the slow flow velocity
of dye in the normal coronary arterial tree may be
related to the increased capillary bed resistance and that
the myocardial dysfunction and enlargement of the heart
represent the aspect of scleroderma cardiomyopathy.

The average survival after the onset of cardiac disease
in scleroderma is 30 months. Our patient has been
known to have cardiomegaly since 1958. This suggests
that the prognosis of scleroderma heart disease may
not be as poor as has been previously reported.

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Surgical Treatment of Leukemic
Involvement of the Mitral Valve*

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A patient with severe mitral insufficiency due to infiltra-
tion of the valve and myocardium by leukemic lympho-
cytes is presented. Treatment was replacement of the
valve with a prosthesis. The significance of this patient
lies in the rarity of the clinical state and the novelty of
treatment.

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LEUKEMIC INVOLVEMENT OF MITRAL VALVE 119
We have recently seen a patient with mitral valvular dysfunction resulting from leukemic infiltration treated by replacing the involved valve. The leukemic infiltration of myocardium caused some of the sutures in the valve annulus to dehiscence producing a para-valvular leak. That tumor-invaded myocardium has less than normal tensile strength is suggested by the occurrence of congestive heart failure1 and by the rare sequela of myocardial rupture.5

Case Report

A 48-year-old black man was admitted to the hospital with increasing dyspnea. Chronic lymphocytic leukemia had been diagnosed three years prior to admission. At the time of diagnosis, he had mild hepatomegaly. A grade 2/6 short early systolic murmur was present along the left sternal border in the supine position, but disappeared in the upright position. This murmur was absent 14 years earlier when he was examined at this hospital. He was asymptomatic and no chemotherapy was given. He was periodically examined in the hematology clinic and no new findings were noted. Three weeks prior to admission the patient reported the onset of dyspnea and two-pillow orthopnea. Physical examination disclosed ventricular extrasystoles, diminished S1, and an apical grade 4/6 pansystolic murmur radiating to the axilla. A diagnosis of mild congestive heart failure, secondary to mitral valvular dysfunction probably of rheumatic origin was made, and the patient was dismissed on furosemide and digoxin.

On admission to the hospital, significant physical findings included diminished S1, loud S3, S4, and early 2/6 diastolic rumbling and a 4/6 pansystolic apical murmur. Leukocyte count was 38,000 with 84 percent lymphocytes. The hematocrit was 41 percent. Prothrombin time was 14.2 with a control of 11.7 seconds. Serum electrolytes, urea nitrogen, glucose and bilirubin were within normal limits. Blood cultures were negative. Electrocardiogram showed frequent, multifocal, sequential premature ventricular beats and high voltage. Chest x-ray film demonstrated borderline cardiomegaly, pulmonary vascular congestion, and right pleural effusion. The patient improved somewhat following drug therapy, but congestive heart failure persisted.

Cardiac catheterization revealed severe mitral regurgitation. The mean pulmonary capillary wedge pressure was 17 mm Hg with a V wave of 34 mm Hg. The mean pulmonary artery pressure was 32 mm Hg and the left ventricular end-diastolic pressure was 20 mm Hg. Mitral valve replacement was decided upon to control the heart failure. At operation the anulus and the mitral leaflets were thickened. Some of the chordae tendineae were fused. A Beall prosthesis was used. Because of the friability of the annular tissue, the mechanism in our case probably of rheumatic origin was made, and the patient was dismissed on furosemide and digoxin.

At autopsy the heart was hypertrophied and dilated. A small infarct of the interventricular septum was noted. There was suture line dehiscence of 5 mm (Fig 1). Microscopic examination of the left atrium and ventricle revealed focal fibrosis associated with dense infiltration of uniform lymphocytes. Sections of the surgically excised mitral valve leaflets and annulus disclosed fibrosis and heavy infiltrate by lymphocytes admixed with fibroblasts and histiocytes. The three other cardiac valves were only moderately infiltrated by leukemic cells and revealed no significant fibrosis. The coronary arteries revealed no tumor infiltration or significant atherosclerosis.

Moderate lymphocytic infiltration of the portal tracts of the liver and of the bone marrow was also noted. Axillary, mediastinal and cervical lymph nodes revealed the sinusoids to be present and dilated. The germinal centers of the lymph nodes were not preserved. Mild lymphocytic infiltration was also noted in the kidneys and alveolar septae of the lungs.

Comments

While the incidence of cardiac metastases in patients dying of unselected malignancies is between 3 percent6 and 21 percent,4 that for leukemic patients is 34 percent5 to 46 percent.6,7 Chronic lymphocytic leukemia was found in one report to involve the heart in 10 of 17 cases.2 Distinctly uncommon is valvular involvement by secondary neoplasms in any of these cases. Roberts7 reported one case of leukemic cell infiltration of the tricuspid valve in a case of acute leukemia, although no cardiac symptoms developed.

Whether cardiac metastases occur most commonly by a hematogenous route,6 or via retrograde flow from involved mediastinal lymph nodes,8 the relative avascularity and the scarcity of lymphatics of the valves provide an anatomic explanation for their immunity. Valvular metastases may also occur by implantation of blood borne cancer cells within the chamber, or by extension from involved myocardium. The mechanism in our case was uncertain. No prior valvular damage could be documented in our case even though one could not exclude prior rheumatic valvulitis.

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Active Pulmonary Hemorrhage Localized by Selective Pulmonary Angiography*

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Massive hemoptysis in a young woman with negative chest film findings is presented. By using selective pulmonary artery angiography during active pulmonary bleeding, the following findings were demonstrated: (1) intraparenchymal hemorrhage, (2) clearing of blood from the lung and bronchial tree by coughing, (3) early filling of the inferior pulmonary vein. Following lobectomy, specimen angiography suggests the presence of a small arteriovenous fistula. This experience demonstrates that selective pulmonary arteriography may be a useful adjunct in the management of selected patients with massive hemoptysis of obscure etiology.

A case of massive hemoptysis in a young woman with a negative chest film is presented. Active pulmonary hemorrhage originating from a small branch of the right lower lobe pulmonary artery was demonstrated by selective pulmonary angiography. Following lobectomy, specimen angiography confirmed the presence of a small arteriovenous communication.

CASE REPORT

A 25-year-old woman was admitted to Prince George's General Hospital on May 23, 1972, because of massive hemoptysis. Except for tachycardia, the physical examination gave negative findings. The admission chest film and a perfusion lung scan were negative. Flexible fiberoptic photobronchoscopy performed during active bleeding revealed blood in every bronchial orifice, even after saline solution irrigations. After three more bleeding episodes within 24 hours, pulmonary angiography was performed revealing a peculiar prominent transverse arterial branch in the right lower lobe. Selective pulmonary arteriography was performed in order to more closely examine this questionable area. This procedure was accompanied by another explosive hemoptysis. The early film (Fig 1) demonstrates contrast material outside the vascular structures appearing within the lung parenchyma. During this injection, at approximately 2 seconds, the patient coughed up blood (containing contrast material), some of which was seen to move within the bronchus. On the 38 second film (Fig 2), the intraparenchymal contrast material has been expelled by the cough into the bronchus. Also demonstrated was the unusual vessel (arteriovenous communication) as well as early filling of the right inferior pulmonary vein.

Following right lower lobectomy, the specimen was injected with contrast material via the right lower lobe artery. The injected specimen (Fig 3) confirms an arteriovenous communication as the inferior pulmonary vein is seen filling from the arterial side via the small aberrant vessel, although the position is slightly distorted due to the contracted state of the specimen.

Two years following surgery the patient has remained asymptomatic.

DISCUSSION

Massive hemoptysis is a life-threatening complication of numerous cardiopulmonary diseases. One of the most difficult problems in pulmonary medicine is the establishment of a cause of massive hemoptysis in the patient with a normal chest film. Specific surgical therapy can only be undertaken with an accurate diagnosis. Bronchoscopy, though often rewarding, during severe hemorrhage may not yield the origin of the bleeding because of rapid flooding of the tracheobronchial tree by fresh blood. With persistent hemorrhage, other diagnostic methods must be performed. Included among these procedures are bronchography, fluoroscopy, scintillation scanning studies, tomography, pulmonary arteriography, selective bronchial arteriography, and in certain instances, cardiac catheterization.

In this particular case, when selective catheterization of the right lower lobe artery was performed, fortuitously the patient bled, thus allowing the serial demonstration of (1) intraparenchymal hemorrhage, (2) clearing of blood from the lung and bronchial tree by coughing, as well as (3) demonstrating clearly early filling of the...