Massive Occlusion of the Main Pulmonary Artery and Primary Branches

Case Report

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This case of massive thrombotic occlusion of the main pulmonary artery is reported because of the size, extent, and duration of the process, because of the prominent findings leading to the correct clinical diagnosis, and, finally, because of the interesting secondary pathologic changes including myocardial infarction without significant coronary arteriosclerosis or occlusion.

Massive occlusion of the main pulmonary artery and its primary branches is believed seldom compatible with prolonged survival. However, a number of reports exist indicating a duration of from several months to several years. No documented cases were found to compare, either in length of life after the episode or in degree of pulmonary artery occlusion, with our patient who survived about 6 years. Chronic obstruction of the major pulmonary arteries with cor pulmonale has been reported as a clinical syndrome more frequently in recent years.1-4 The cause of pulmonary embolism seems directly related to peripheral vascular disease, namely, phlebothrombosis or possibly thrombophlebitis. Emboli are considered to be uncommon as a result of thrombophlebitis, but a red, swollen, tender extremity may represent phlebothrombosis rather than phlebitis, thus causing a misleading opinion regarding the origin of emboli and erroneously raising the frequency of embolism reported as due to thrombophlebitis. The total number of reported cases of long-standing pulmonary embolism may be less than 200 and the majority of these has been discovered at autopsy. No reports were found in which an antemortem diagnosis was made before 1940.

This report is intended to call attention to 2 important phases of this condition. First the duration of life after the initial episode (1950) or following ones (1951), (at least 4 and possibly 6) and second, the recognition of long-standing thromboembolic occlusions of major pulmonary arteries.

Patients surviving the acute shock of large pulmonary emboli will benefit from present-day cardiovascular surgical procedures and medical therapy. Severe pulmonary hypertension and its concomitant right heart failure may be prevented or altered. The procedures of embolectomy, endarterectomy, and artery resection will have their place for certain patients. Indeed, one report is available from France of a cure by resection of the involved segment of pulmonary artery.5 The use of anticoagulant drugs is restricted to the prevention of the primary thrombosis; they cannot dissolve thrombi or emboli. However, they may reduce embolization by inhibition of further clotting.6

It is important to emphasize that patients may survive after severe embolization for a prolonged period as shown by the case to be presented. The long duration of symptoms is not at all incompatible with a clinical diagnosis, which should be suspected, particularly in younger patients, with bizarre symptoms related to pulmonary and cardiovascular systems. In part, some of the reasons for infrequent or belated diagnosis may be due to adherence to too rigid criteria, with classical symptoms and signs developing only in the late phases of pulmonary hypertension.7-8 Cardiac catheterization and angiocardiography are useful special diagnostic procedures, but the diagnosis of obstruction of the major pulmonary arteries should be suspected or estab-
lished on firm clinical grounds prior to the use of these procedures.

Symptoms and signs of pulmonary embolism include shock, syncope, weakness, cough, dyspnea, sweating, chest pain, and cardiac arrhythmia; later, cyanosis, hemoptysis, clubbing of digits (if arterial oxygen saturation is chronically less than 70 to 88 per cent), and hoarseness may be seen. Right heart dilatation (cor pulmonale) and failure occur secondary to pulmonary hypertension. X-ray studies may reveal a hilar mass or prominent vascular patterns and clear pulmonary fields; or cardiac enlargement may be present. The left hilar shadow consists of the main pulmonary artery and proximal portion of the left main branch, so that if these are occluded, fluoroscopy will not show hilar and peripheral vascular hyperactivity or “dance.”9 Murmurs are not characteristic and may be due to turbulence in the pulmonary artery from the clot. Electrocardiographic evidence of right ventricular hypertrophy or strain often indicates a pulmonary artery pressure greater than 30 mm. Hg and signifies far advanced disease.10, 11

The anatomic components of pulmonary hypertension include thrombi of varying stages and vascular intimal and medial thickening; possibly necrobiotic constriction of pulmonary vessels is a physiologic component.

**Pathology**

Pathologic changes resulting from long-standing massive thrombotic occlusions of pulmonary arteries usually include hypertrophy and dilatation of the right side of the heart. The degree of organization and recanalization of the thrombotic emboli is dependent upon time and the sizes of the vessel involved and of the clot itself; many thrombi of long standing in larger vessels show almost complete lack of organization and are only a densely hyalinized mass. Arteriosclerosis of major and minor pulmonary arteries seems to result mainly from the incorporation of thrombi in the vessel wall. All degrees of change can be seen in the lung itself from a completely normal microscopic picture through minor changes as a result of old infarction, to grossly and microscopically obvious healed infarcts. Secondary inflammatory processes, of course, are frequent.12, 13

**Case Report**

L. J., a 40-year-old white man was first seen early in 1955, complaining of shortness of breath, palpitations, loss of strength, and loss of 15 pounds in weight; examination revealed cyanosis, early clubbing of fingers and toes, hypotension (100/70), cardiac enlargement, pulmonary congestion, polycythemia, a cold left leg, and a superficial “thrombophlebitis” of the right calf. There was a grade II, apical, systolic murmur and the pulmonic second sound was duplicated. Lower sternal dullness was noted. The electrocardiogram indicated right ventricular hypertrophy and incomplete right bundle-branch block (fig. 1).

He gave a history of varicosities of both legs and “phlebitis,” (undocumented as to thrombophlebitis) beginning in 1943, and resulting in a varicose ulcer on the left leg by 1946, for which a left superficial saphenous ligation was done in 1947. In early 1950, he suffered sudden chest pain and hemoptysis and later had 5 or 6 similar attacks in 1951. At this time he was given digitalis because of extrasystoles. In October 1954, recurrent ulcers of the left leg were treated by stripping of an upper leg vein. The cardiopulmonary disease and syndrome of pulmonary artery obstruction remained undetected although a chest x-ray as early as 1951 had revealed pulmonary densities and prominent hilar shadows (fig. 2). Blood pressure varied between 80/50 and 100/70. Prior to death in March 1956, the hemoglobin was 18.2 Gm. per 100 ml. and the hematocrit value was 59.

**Autopsy Findings.** The main pathologic finding was a single, huge, old, thrombotic pulmonary embolus involving the main pulmonary artery and
Fig. 2. Progressive cardiac enlargement, hilar prominence, and clear lung fields, beginning in 1951, to pulmonary nodularity in 1956.

Fig. 3. Longitudinal section of extrapulmonary portions of main pulmonary artery and right and left branches; the main artery is below; the longer limb is the right branch and it shows more recent thrombosis. The wooden probes indicate the remaining channels to the right and left sides.

its major branches bilaterally (fig. 3); the proximal end was just 1 cm. above the pulmonic valve ring. This artery and its branches were longitudinally sectioned and were estimated to be, in any single cross-sectioned area, at least 90 per cent filled with thrombus, which was old and laminated for the most part, pale, and firm (fig. 4). At a few points, especially in the distal segment of the right main arterial branch, there were softening, friability, and roughening, suggesting that some portions here may have been more recent. When this mass was followed out into the pulmonary arterial branches on both sides, numerous and irregular extensions into the smaller branches were revealed. A very tortuous channel around the thrombus existed from the proximal portion of the pulmonary artery into the periphery of both lungs. In the more peripheral portions of the arterial tree numerous other thrombotic masses were apparent. Some of these were organized and at least 1, measuring 6 mm. in diameter and progressing for a distance of approximately 2 cm., was not connected with the main thrombus.

There were marked right ventricular and right atrial hypertrophy and dilatation (fig. 5). The heart itself weighed, after fixation, 635 Gm. and measured 12.5 cm. in maximum transverse diameter.

There were 2 old, well healed myocardial infarcts and widespread myocardial fibrosis, without demonstrable coronary artery disease. One of these infarcts in the inferior portion of the interventricular septum extended to involve the right ventricular endocardium. Chronic progressive mural thrombosis of the right ventricle overlay this infarct. Another old thrombus was present in the right atrial appendage.
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The coronary arteries were thin-walled and elastic, showing no arteriosclerosis. The pulmonic valve ring measured 9.3 cm. in circumference; other valvular measurements were normal and there was no other significant valvular distortion. There were chronic focal degenerative pulmonary changes, possibly representing old, healed, patchy pulmonary infarcts. Chronic passive congestion of the liver was observed.

An acute suppurative bronchitis with minimal focal bronchopneumonia was also present.

DISCUSSION

Clinicopathologic Correlation. Apparently this man suffered a tremendous pulmonary embolism in 1950, 6 years before death, and the
majority if not all of the disease resulted from this. There were other similar clinical episodes in 1951, probably as a result of smaller emboli breaking off from the major mass in the pulmonary artery, or possibly as a result of additional thrombosis being superimposed upon the original embolus, as suggested by the lamination. Obstruction of the pulmonary arterial system remained undetected through 1954. When seen early in 1955, he had definite evidence of severe chronic cor pulmonale by physical examination, x-ray, and electrocardiographic study.

The 2 old myocardial infarcts and focal myocardial fibrosis cannot be explained as the result of coronary artery disease; we think these have resulted from anoxemia due to pulmonary vascular obstruction. The right ventricular thrombosis was thought to be secondary to the myocardial infarction; it is rare for thrombosis to occur in the right ventricle from any cause, and therefore its occurrence here is probably connected with cor pulmonale. The thrombus in the right atrial appendage is readily explicable by right heart failure, as is the congestion of the liver.

**Summary**

A case of thrombotic pulmonary embolism is reported. This is apparently the most massive pulmonary artery obstruction of any significant duration in the literature; the length of life after the initial episode (6 years) is therefore especially surprising and also the longest yet reported.

A plea is made for proper etiologic diagnosis of cor pulmonale as well as for recognition of the syndrome of chronic pulmonary vascular obstruction.

**Summario in Interlingua**

Es reportate un caso de thrombotic embolismo pulmonar. Isto es apparentemente le plus massive obstruction pulmono-arterial de duratión significative in le litteratura. Le superventia durante 6 annos post le episodio initial es specialmente surprendeante. Illo es le plus longe unquam reportate.

Es signalate le urgenta necessitate del correcte diagnose etiologic de corde pulmonale e del recognition del syndrome de chronic obstruction pulmono-vascular.

**REFERENCES**

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