Observations on the Natural History of Varicosity of Pulmonary Veins

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SUMMARY

Pulmonary vein varix was diagnosed by roentgen studies in three patients. One showed enlargement of the lesion in 7 years with progression of heart disease and regression of the varix after correction of the mitral insufficiency. The other two patients without associated heart disease demonstrated no change in the varicosity of the pulmonary vein after 4 and 15 years.

ADDITIONAL INDEXING WORDS:

Mitral insufficiency
Valsalva maneuver
Müller maneuver
Radiological examinations
Angiocardiography

PREVIOUS reports have alluded to the rarity of varicosities of the pulmonary veins. To date, only 18 cases exist in the literature. Six were diagnosed at autopsy1–6 and the rest by radiological methods.7–15

Little is known about the course and outcome of pulmonary venous varices. The present communication is a report of three cases. In one patient, the lesion enlarged in 7 years with progression of mitral insufficiency and then disappeared 2 years after prosthetic replacement of the patient's mitral valve. The other two patients were asymptomatic and showed no change in the state of the varix for 4 and 15 years.

Report of Cases

Case 1 (GNHCH 50-73-36)

A white woman, 33 years old, had symptomatic cardiac disease with features by history and physical and laboratory examinations consistent with rheumatic mitral insufficiency and recurrent congestive failure.

Cardiac Catheterization

The major findings were hypertension in the right ventricle (45/3 mm Hg) and left atrium (v wave = 35 mm Hg). The left ventricular pressure was 85/10 mm Hg.

Radiological Examination

Chest x-rays with barium swallow revealed massive cardiomegaly with predominant enlargement of the left heart. There was a teardrop density at the right paracardiac area just above the right hemidiaphragm (fig. 1 A). On fluoroscopy, no intracardiac calcifications were seen. There were no pulsations in the right paracardiac mass. The Valsalva maneuver decreased and the Müller maneuver increased the size of the mass. Laminagrams confirmed the observation that the mass was noncardiac and without calcifications (fig. 2). Comparison with x-ray studies carried out 7 years previously revealed slight enlargement of the right paracardiac mass and cardiomegaly.

Right atrial angiocardiography demonstrated minimal enlargement of the right atrium and ventricle. The teardrop density in the right paracardiac area opacified in the sequential pulmonary venous phase (fig. 3 A and B). The density remained even though the normal veins were already clear of the contrast material. The left atrium and ventricle were greatly dilated. The circulation was slow without evidence of a shunt. There was poor egress of radiopaque material into the normal-sized aorta.

Surgery

Cardiac exploration revealed a jet lesion in the massively dilated left atrium. The anterior leaflet of the mitral valve was redundant and overriding. The posterior leaflet was shortened with
Chest roentgenogram in case 1. (A) Cardiomegaly and right paracardiac mass. The mass decreased in size with a Valsalva maneuver and enlarged with a Muller maneuver during fluoroscopy. (B) Two years after cardiac surgery. The heart has diminished slightly in size and the pulmonary vein varix has disappeared.

**Figure 2**

Laminagram demonstrates the paracardiac mass in case 1 to better advantage than figure 1 A.

distorted rolled edges. The mitral valve, including the chordae tendineae, was excised and a prosthetic Starr valve was inserted. The incision was closed without further exploration of the right paracardiac pulmonary vein varix.

**Progress**

For 2 years after surgery, the patient progressively improved. Her last chest x-rays showed reduction in the cardiomegaly and disappearance of the pulmonary vein varix (fig. 1 B).

**Case 2 (GNHCH B8-82-84)**

An asymptomatic, 21-year-old, colored woman was admitted for evaluation of abnormal chest x-rays. Her first chest x-rays, made 2 years before, showed a left hilar mass. Her father died of pulmonary tuberculosis 8 years previously. For these reasons, she was immediately started on antituberculous therapy.

There was no abnormal physical finding. Likewise, the laboratory examinations were noncontributory.

**Radiological Examinations**

The chest roentgenograms and laminagrams (fig. 4 A and B) demonstrated a lobulated finger-like density posterior and inferior to the bifurcation of the left bronchus, directed to and in the vicinity of the left atrium. Valsalva maneuvers during laminography failed to show change in the size of the mass.

Angiography (intravenous and selective pulmonary arteriograms, fig. 4 C) demonstrated the density to be a dilated pulmonary vein of one of the superior venous trunks. The rest of the angiograms were normal.

**Progress**

Antituberculous treatment was immediately stopped when the diagnosis was made. The lesion did not change in 4 years.

**Case 3 (J.D.)**

An asymptomatic, 37-year-old, white man was admitted to Bridgeport Hospital for evaluation...
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Figure 3
Pulmonary arteriographic phase (A) shows lack of filling of the mass which opacified in the pulmonary venous phase (B [right]). There is stagnation of contrast material in the varix as compared to the rest of the pulmonary veins.

Figure 4
Chest x-ray (A) in case 2 shows a left hilar mass. Additional studies by laminography (B) and selective pulmonary arteriography (C) reveal the location and nature of the mass—pulmonary vein varix.

of the density in the right middle lobe of the lung. Fifteen years prior to admission a density in the right lung was observed in a routine chest x-ray. Annual x-rays complemented by additional examinations showed no change.

Physical and laboratory examinations were negative. Right heart catheterization gave normal results.

Radiological Examinations
Chest x-rays showed a small nodular density in the right perihilar area (fig. 5 A). Minimal pulsations were observed at the border of the mass on fluoroscopy. There was no significant change with the Valsalva or Müller maneuvers. Angiocardiography demonstrated the pulmonary varix, opacifying during the normal venous phase (fig. 5 B).

Discussion
Varicosity of the pulmonary vein can be seen during thoracotomy and postmortem examinations. Clinically there are no signs or symptoms directly referable to the lesion. However, pulmonary varices contributed to
death in three patients whose cases are reported in the literature: apparent bleeding into a bronchus,\(^2\) rupture of the varix,\(^3\) and cerebral embolism.\(^4\)

The diagnosis is essentially a radiological one. The concern of the physician over a lobulated mass or masses in chest roentgenograms leads to further investigation. Different maneuvers during fluoroscopy may aid in the identification of a vascular mass (case 1). The varix becomes small with the Valsalva maneuver and enlarges with the Müller maneuver.\(^1\)\(^1\) Cases 2 and 3, however, showed no change with these maneuvers. Laminagraphy has been used also in demonstrating the lesion.\(^7\)\(^-\)\(^9\)\(^,\)\(^1\)\(^5\)

The definitive diagnosis is made by angiocardiography.\(^1\)\(^0\)\^-\(^1\)\(^6\) The lesion opacifies during the pulmonary venous phase. There is stasis of radiopaque material in the varix.\(^1\)\(^5\) It has no abnormal arterial communications. If a filling defect is seen, it indicates thrombosis in the varix which has been found only at autopsy.

Several authors\(^3\)\(^,\)\(^6\)\(^,\)\(^1\)\(^1\)\(^,\)\(^1\)\(^2\)\(^,\)\(^1\)\(^4\)\(^,\)\(^1\)\(^7\)\(^,\)\(^1\)\(^8\) reported localized or generalized dilatation of pulmonary veins in certain congenital and acquired heart diseases. Klinck and Hunt\(^6\) theorized that the varices are congenital, and probably insignificant until concomitant disease leads to increasing phlebectasia. In support of this theory, case 1 showed enlargement of the varix in 7 years with progression of the mitral disease. Spontaneous regression of the varix was observed in 2 years after correction of the mitral insufficiency.

The lesions were isolated and insignificant in the other two patients. The varices showed no change after 4 and 15 years. This is similar to the case reported by Gottesman and Weinstein\(^1\)\(^1\) which demonstrated no change 10 years after mitral commissurotomy.

Figure 5

*Figure 5*

*Chest x-ray (A) in case 3 reveals a nodular mass in the right perihilar area. On angiography (B) this mass proved to be a tortuous and dilated pulmonary vein.*
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References

Responsibility

SOCRATES: Well, look here. Suppose someone went up to your friend Eryximachus, or his father Acumenus, and said, 'I know how to apply such treatment to a patient's body as will induce warmth or coolness, as I choose; I can make him vomit, if I see fit, or go to stool, and so on and so forth. And on the strength of this knowledge I claim to be a competent physician, and to make a competent physician of anyone to whom I communicate this knowledge.' What do you imagine they would have to say to that?

PHAEDRUS: They would ask him, of course, whether he also knew which patients ought to be given the various treatments, and when, and for how long.

SOCRATES: Then what if he said, 'Oh, no, but I expect my pupils to manage what you refer to by themselves?'

PHAEDRUS: I expect they would say, 'The man is mad; he thinks he has made himself a doctor by picking up something out of a book, or coming across some comon drug or other, without any real knowledge of medicine.'—EDITH HAMILTON and HUNTINGTON CAIRNS (Ed.): The Collected Dialogues of Plato. New York, Bollingen Foundation (Pantheon Books; Bollingen Series 71) 1961, p. 513.
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