Abnormal Pulmonary Vessels in a Patient with Chest Pain

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A 74-year-old white woman entered University Hospital with right pleuritic chest pain, dyspnea, and right calf pain 11 days after urethral dilatation incident to recurrent urinary tract infections.

Physical examination disclosed a few basilar rales bilaterally and slight right calf tenderness. There were no apparent telangiectases. The chest roentgenograms showed no abnormalities. Right heart strain was indicated by electrocardiogram. Pulmonary perfusion scan demonstrated a perfusion defect at the right base, while the xenon ventilation scan was negative. Blood gas values: $P_{O_2}$ 87, $P_{CO_2}$ 20, serum bicarbonate of 13 mEq/L and pH 7.43; hemoglobin and hematocrit were 12.6 grams and 37 percent respectively. At pulmonary arteriography a normal main pulmonary artery pressure (30/8 with a mean of 15 mm Hg) was recorded.
Diagnosis: Minute Pulmonary Arteriovenous Fistula with Anomalous Pulmonary Venous Drainage

As is frequently the case, small vessel communications between the pulmonary artery and vein may not actually be seen. The clue then to the diagnosis is the early opacification of the venous circulation. Contrast material appears in the innominate vein immediately following pulmonary artery injection by means of an anomalous distribution of left upper lobe venous drainage (Fig 1 and 2). The fistula, in this case, is situated in the left upper lobe and the distribution of the apical-posterior branches of the pulmonary artery.

Microscopic arteriovenous shunts in normal lung of the order of 35 to 200 microns have been demonstrated by several investigators.1 Gas experiments showing venous admixture in the absence of intracardiac shunts along with deployment of glass beads in pulmonary arterial obliterative disease substantiate the presence of pulmonary microscopic arteriovenous shunts. These shunts apparently may enlarge when the pulmonary arterial systolic pressure exceeds 40 mm Hg.2 There has been no convincing demonstration of these physiologic shunts by in vivo angiography in the human lung. Magnification studies have demonstrated, on the other hand, minute abnormal fistulas of the order of 500 microns to 2 mm.3

Abnormal congenital shunts have been classified by Anabtawi, Ellison, and Ellison4 into five categories based upon embryologic possibilities:
1. Multiple small A-V fistulas without aneurysm
2. Large A-V aneurysm (peripheral)
3. A) Large A-V aneurysm (central)
3. B) Large A-V aneurysm with anomalous venous drainage
3. C) Multiple small A-V fistulas with anomalous venous drainage
4. A) Large venous aneurysm with systemic artery communication
4. B) Large venous aneurysm without fistula
5. Anomalous venous drainage without fistulas

Pulmonary arteriovenous fistulas may be congenital or less commonly acquired. The latter have been known to occur in hepatic cirrhosis, pulmonary schistosomiasis and metastatic thyroid carcinoma. In addition, bronchial-to-pulmonary artery shunts sometimes develop in chronic lung states such as bronchiectasis and bronchial to pulmonary vein shunts in advanced emphysema. Congenital fistulas occur singly in about two-thirds of cases and are most commonly located in the lower lobes (right more often than left). Rendu-Osler-Weber's hereditary hemorrhagic telangiectasia is reported in 40 to 65 percent of patients with congenital arteriovenous fistulas in the lung. There was no evidence of this in the present case which, though single, corresponds with type 3 C of Anabtawi, Ellison and Ellison based upon embryologic possibilities.

An isolated minute pulmonary arteriovenous fistula even without anomalous venous drainage is, of course, of doubtful hemodynamic significance in the presence of normal pulmonary arterial pressure. However, pulmonary arterial hypertension may effect relative hypoxia by enlargement of normally present shunts (those less than 200 microns). A similar situation may exist in the presence of atelectasis or multiple small thromboemboli where we have sometimes observed early venous opacification at pulmonary arteriography. With multiple abnormal fistulas, even more significant hypoxia seems likely to result.

REFERENCES