Lung Tumor in a Patient with Congenital Unilateral Hypoplasia of the Pulmonary Artery*

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The occurrence of an undifferentiated carcinoma in the affected lung of a cigarette smoker with asymptomatic congenital hypoplasia of the right pulmonary artery is reported.

Reports of congenital aplasia or hypoplasia of a main branch of the pulmonary artery have appeared in the medical literature since 1868. Dotter and Steinberg in 1949 reported a case of hypoplastic pulmonary artery secondary to widespread unilateral lung disease. In 1953, Swyer and James reported a patient with congenital abnormality of the pulmonary artery and unilateral emphysema. Since then, a number of similar cases have been described, particularly in the British literature. Congenital unilateral hypoplasia of the pulmonary artery was first documented in life by angiography in 1952 by Madoff et al and later was also observed by others. This syndrome has been associated with tetralogy of Fallot, varices of pulmonary veins draining the opposite lung, and unilateral pulmonary edema. The occurrence of a malignant neoplasm in the hypoplastic lung has, to our knowledge, not yet been reported. The purpose of this report is to present such a case.

Case Report

A 49-year-old asymptomatic black woman was admitted to Mount Sinai Medical Center, Miami Beach, Fla, because of an abnormal preemployment chest roentgenogram. She had an uncomplicated birth and developmental history but remembered occasional shortness of breath while doing exercises in school. The patient denied other respiratory symptoms. At age 28 years, she was told that her heart was “pushed to the right.” She admitted to smoking one pack of cigarettes a day for the last ten years. A previous chest roentgenogram taken nine months prior to the present admission did not show any lesions. Physical examination revealed a slightly obese, normally developed woman in no acute distress. Abnormal physical findings were limited to slight hyperresonance to percussion and distant breath sounds over the right hemithorax. Chest roentgenograms (conventional posteroanterior and lateral projections) revealed volume loss of the right lung with mediastinal shift to the same side and hyperlucency of the left lung. The pulmonary artery segment was not identified on the right side. The heart was not enlarged and had an unremarkable configuration. An ill-defined nodular lesion measuring 1.5 x 1 cm was noted in the anterior segment of the right upper lobe. No hilar lymphadenopathy was detectable (Fig 1). Whole-chest tomograms did not show any calcification or caviation of the lung lesion, nor were satellite lesions or other pulmonary infiltrates seen. A lung scan showed no uptake of technetium-labeled macroaggregated albumin in the right lung. In contrast, the left lung had a homogeneous uptake and appeared slightly enlarged. Xenon ventilation scans revealed a delayed washout on the affected side. A pulmonary angio- gram confirmed the presence of a markedly hypoplastic right pulmonary artery with a prominent pulmonary arterial tree and lower-lobe varix on the left (Fig 2).

Bronchoscopy revealed normal tracheobronchial anatomy with diffuse inflammatory changes of the mucosa. No endobronchial lesion was seen. Because of the enlarged bronchial arterial circulation known to be present in patients with

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hypoplastic pulmonary artery,\textsuperscript{10} no transbronchial biopsy was attempted. Preoperative mediastinoscopy disclosed enlarged anthracotic lymph nodes in the mediastinum which contained undifferentiated metastatic carcinoma.

**DISCUSSION**

Congenital aplasia or hypoplasia of a main branch of the pulmonary artery is a rare malformation. Conventional chest roentgenograms reveal a loss of volume and an absent pulmonary artery segment on the affected side. The diagnosis is confirmed by pulmonary arteriography. The abnormality may occur alone, as in our patient, but it is frequently related to other congenital malformations, especially of the heart and great vessels.\textsuperscript{11} In the absence of associated cardiac malformations, it is usually the right pulmonary artery which is affected.\textsuperscript{12} The combination of hypoplasia of the right pulmonary artery and varices of the veins from the left lung, which was demonstrated in our patient, has also been reported in the past.\textsuperscript{13}

The occurrence of a malignant tumor in the affected lung of a patient with hypoplastic pulmonary artery represents a certain curiosity. Although the true nature of the lung lesion was not determined by thoracotomy or lung biopsy, the presence of metastatic undifferentiated carcinoma in mediastinal lymph nodes in a cigarette smoker made the diagnosis of a malignant lung tumor highly probable. Tumor obstruction of a pulmonary artery is occasionally found with bronchogenic carcinoma,\textsuperscript{11} but the occurrence of carcinoma in a congenital hypoplasia of the pulmonary artery has, to our knowledge, never been reported. Whether the presence of impaired ventilation of the affected lung, which was suggested by the xenon ventilation scan, favored or delayed the development of the carcinoma in this smoker remains an interesting unanswered question.

**REFERENCES**

2. Dotter CT, Steinberg I: Angiographic study of pulmonary artery. JAMA 139:566–572, 1949