Systemic Arterialization of Lung without Sequestration

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Anomalous systemic arterial supply to the lungs has been described in several congenital pathologic entities: bronchopulmonary sequestration, the scimitar syndrome, and anomalous systemic supply without sequestration. The latter is the rarest form of aberrant systemic blood supply to the lung; only four cases have been described in the English literature [1–4]. We report a fifth case of this variety and discuss the spectrum of anomalous pulmonary blood supply.

Case Report

A 26-year-old woman had worsening dyspnea on exertion and recurrent hemoptysis. Four years before, she had had some right-sided pleuritic pain and hemoptysis and was taking oral contraceptives. A chest radiograph at another hospital was interpreted as "normal" and a ventilation-perfusion lung scan revealed a normally ventilated but nonperfused segment in the right lower lobe. Pulmonary angiography was not performed. She was thought to have had an acute pulmonary embolism and was begun on anticoagulants, which were stopped a short time later due to recurrent hemoptysis.

Dyspnea on exertion and recurrent febrile episodes with cough and hemoptysis thought to be upper respiratory infections developed 1 year later. A poorly defined infiltrate in the right lower lobe was identified on chest radiography. Bronchoscopy and bronchography were performed and "bronchiectasis" was diagnosed. Recurrent febrile episodes, pleuritic pain, and hemoptysis occurred with deterioration in her exercise capacity. Bronchoscopy was repeated 3 years later and resection of the right lower lobe was recommended for "worsening bronchiectasis."

Subsequent radiographic evaluation at our hospital demonstrated a patchy area of consolidation in the right lower lobe and a "bandlike" density paralleling the right heart border and extending below the right hemidiaphragm (fig. 1A). This was felt to be a vascular structure and a preliminary diagnosis of the scimitar syndrome was made. She was evaluated by bronchoscopy, pulmonary arteriography, thoracic and abdominal aortography, computed tomography (CT), and ventilation-perfusion lung scan.

Bronchoscopy failed to show any abnormality in the right lung and the right tracheobronchial tree was reported as normal in size, configuration, and position. Pulmonary arteriography (fig. 1B) demonstrated the absence of the normal branches to the basilar segments of the right lower lobe. The rest of the pulmonary arterial tree appeared intact. Normal pulmonary artery pressures were obtained and there was no evidence of a left to right shunt. An aortogram demonstrated a large anomalous artery arising from the celiac axis which supplied the right lower lobe. Venous drainage from the involved segments was to the left atrium (figs. 1C–1E). The anomalous vessel was easily demonstrated by CT using bolus contrast enhancement with scanning timed to the arterial phase (fig. 1F). This vessel was located posterior to the right lobe of the liver adjacent to the inferior vena cava. Ventilation-perfusion lung scan showed a significant ventilation perfusion mismatch in the right lung, the right lower lobe being nonperfused but ventilated relatively normally (fig. 2).

Thoracotomy with ligation of the anomalous vessel and resection of the right lower lobe was later performed. The anomalous vessel was 1 cm in diameter, elastic in type, and coursed through the right lower lobe. An atretic pulmonary artery segment was identified. On pathologic examination, the specimen showed an intact right lower lobe bronchial tree, multiple areas of recent hemorrhage, and scattered hemosiderin-laden macrophages in the parenchyma. There was no evidence of bronchiectasis or chronic infection.

Discussion

In our case, a mistaken diagnosis of pulmonary embolism was initially made because of the suggestive presenting symptoms and ventilation-perfusion lung scans compatible with this diagnosis. This error could have been avoided had the abnormal vascular shadow been recognized. Furthermore, stability of a perfusion abnormality over a period of years is incompatible with pulmonary embolism. Why the diagnosis of bronchiectasis was made remains unclear. Findings of bronchiectasis were present neither on repeat bronchoscopy nor on pathologic examination. Retrospectively, the recurrent episodes of fever, hemoptysis, and cough were due to recurrent pulmonary hemorrhage in the involved segment of lung. Since surgery, the patient has done well and has had no recurrence of her symptoms.

The *sequestration complex*, as first described by Pryce et al. [2], comprises a number of overlapping clinical, radiographic, and pathologic entities whose common denominator is an anomalous systemic arterial supply to the lung parenchyma and atresia or hypoplasia of the pulmonary

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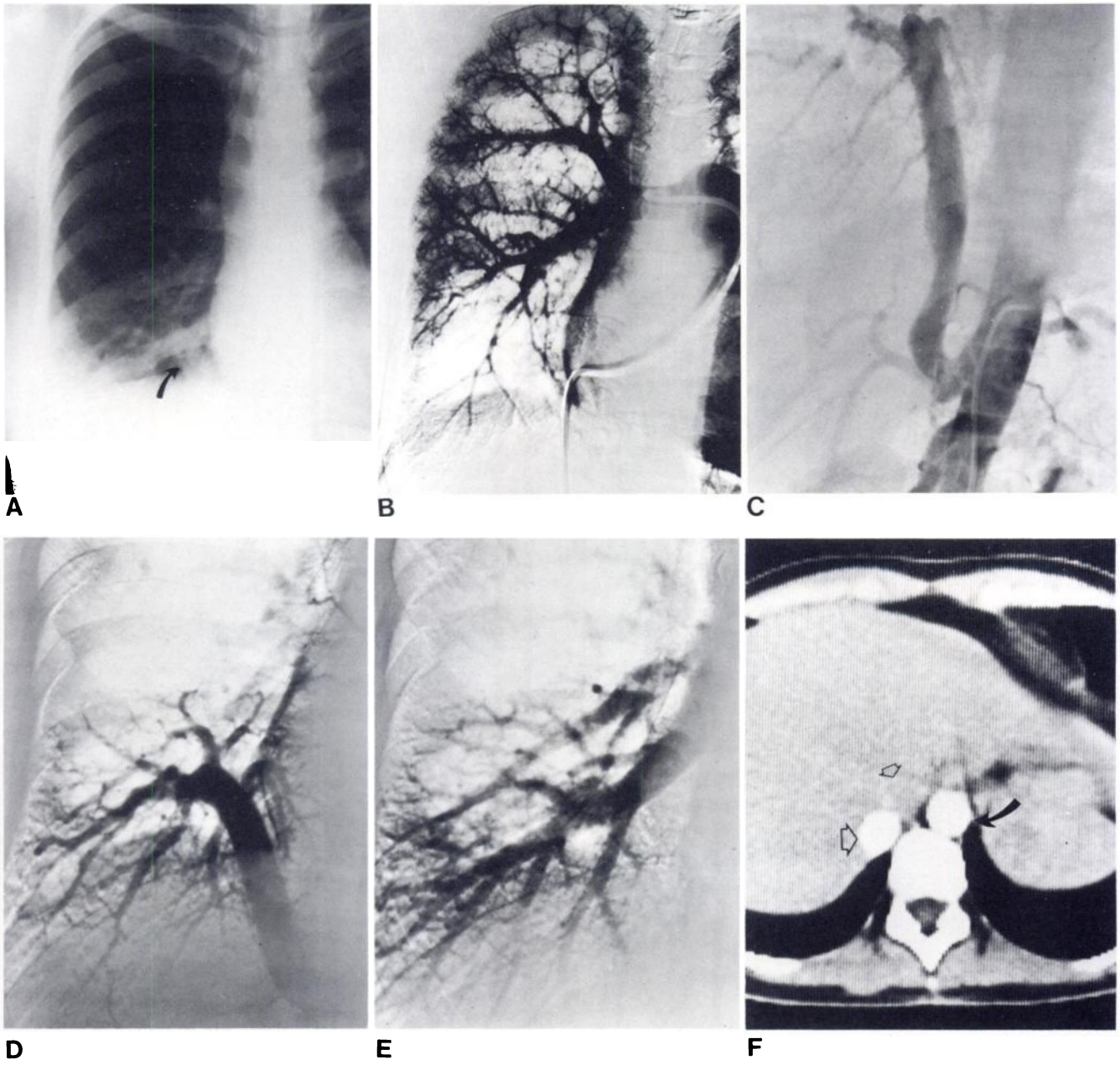
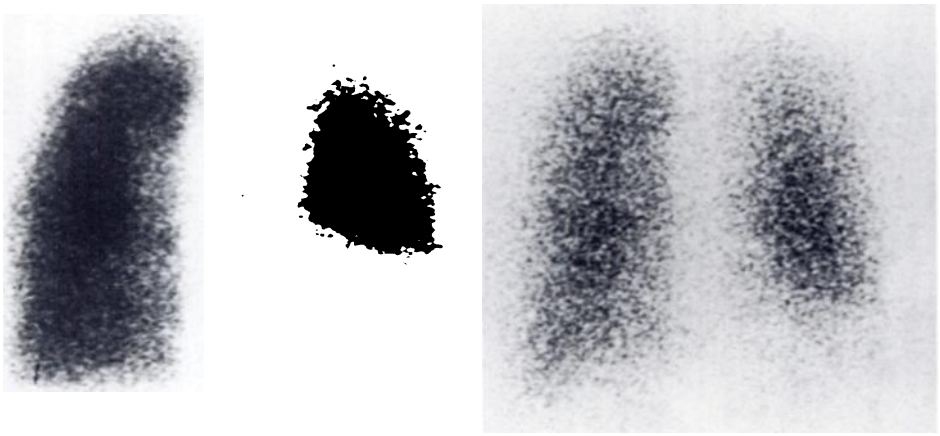


Fig. 1.—**A**, Bandlike density (*arrow*) adjacent to right heart border and patchy consolidation in right lower lobe. Pulmonary artery in right hilum looks smaller than left. **B**, Right pulmonary arteriogram. Branches to basal segments of right lower lobe are absent. **C**, Flush aortogram, right posterior oblique projection. Artery arises from celiac artery. **D**, Selective injection of aberrant artery. **E**, Venous phase. Drainage directly to left atrium. Caliber of vessels suggests high volume flow. **F**, CT scan. Anomalous artery (*large open arrow*) passes through diaphragm. Inferior vena cava (*small open arrow*) and aorta (*solid arrow*).

artery. Other features include cystic bronchial changes, anomalous pulmonary venous return, and foregut anomalies of varying degrees in each specific syndrome. Clinically, however, the entire complex has a similar presentation including an asymptomatic radiographic abnormality, hemoptysis, and/or recurrent pulmonary infection.

The most frequently recognized form of the "sequestration complex" is bronchopulmonary sequestration, a congenital lung malformation in which a part of the lung, most often a lower lobe segment, is completely (extralobar) or incompletely (intralobar) separated from normally functioning lung tissue. By definition, the sequestered lung receives its arterial blood supply from an aberrant artery arising from the aorta. Intralobar sequestrations are contained within the normal pleural cavity while the extralobar variety is enveloped in its own distinct pleura. Venous drainage may be entirely normal or completely anomalous. A spectrum of anomalous connections with the foregut has been reported with the extralobar type [5]. An inherent part of this malformation, as classically described, is the absence



A B

Fig. 2.—**A**, ^{99m}Tc perfusion lung scan, posterior projection. Perfusion of right lower lobe basal segments is lacking. **B**, ^{133}Xe ventilation lung scan, posterior projection. Ventilation of right lower lobe present but minimally decreased.

of communication between the normal bronchial tree and the sequestered segment. Cystic, air-containing areas present in the abnormal segment have been ascribed to collateral alveolar ventilation or infection with secondary fistulous communication with the bronchial tree. Bronchography typically shows displacement of the normal bronchial tree by the sequestered segment. Several investigators have demonstrated bronchographic filling of the cystic spaces [6–8], presumably through fistulae [9].

More recently, Takahoshi et al. [6] and Groot [10] demonstrated that the communication between sequestered lung and bronchial tree may contain normal continuity of mucosal and submucosal bronchial elements without intervening inflammatory changes. Thus, it appears that bronchial isolation is not universal in sequestration.

The scimitar syndrome consists of anomalous venous return from the right lower lobe, usually draining to the inferior vena cava, portal vein, a hepatic vein, and, rarely, directly to the right atrium. Systemic arterial supply is a frequent but inconstant finding and, when present, is accompanied by varying degrees of pulmonary artery hypoplasia. While lobar agenesis most often occurs in this syndrome, normal bronchial anatomy and sequestration have been reported [9].

The rarest form of anomalous systemic arterial supply to the lung is the type described here, in which an anomalous systemic artery supplies a normal unsequestered segment of lung. The four reported cases have involved the right lower lobe, and, in all cases, the systemic artery arose from the proximal abdominal aorta or the celiac axis. Anomalous venous return has not been reported in this syndrome, while atresia of the corresponding pulmonary artery segment was always present. In effect, the anomaly is a left-to-left shunt.

Rarely, acquired disorders of the lung are seen with systemic arterialization of the lung parenchyma. Whenever there is chronic oligemia, systemic collaterals from bronchial, intercostal, internal thoracic, or phrenic arteries may develop. Patients with marked pleural adhesions can also develop shunting from the high pressure circulation of the parietal pleural to the low pressure system of the pulmonary bed in the absence of oligemia. In both cases, collateral vessels are seldom visible on radiographs.

During embryonic development, the lung receives a dual blood supply from the aorta consisting of bronchial and splanchnic vessels. The splanchnic supply normally regresses once the pulmonary arterial system becomes established, but may persist to varying degrees if this normal course of events is interrupted. In all settings except pleural adhesions, pulmonary oligemia is a constant underlying factor with either persistence or reestablishment of pulmonary-splanchnic collaterals.

To summarize, persistence of the embryologic circulation of the lung is most often associated with an intralobar sequestered lung with anomalous systemic arterial supply and pulmonary artery atresia. However, a wide spectrum of abnormalities can be seen and is probably best termed the *sequestration complex*, as first described by Pryce et al. [2]. Our case represents an example of the rarest form of this complex, anomalous systemic arterial supply without pulmonary sequestration.

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