Hypoxemia in Lipoid Pneumonia: Role of Intrapulmonary Bronchopulmonary Anastomoses

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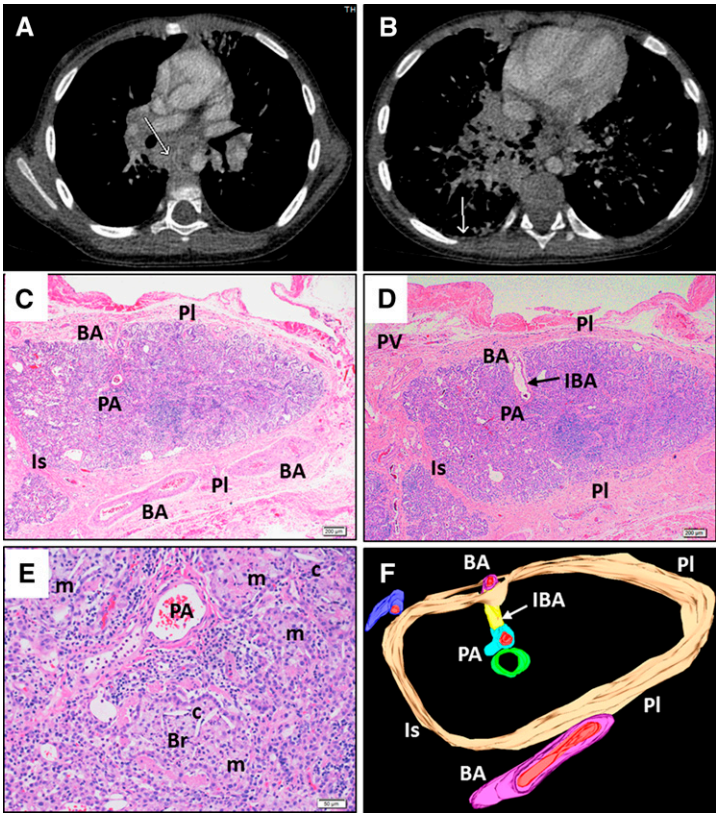

Figure 1. (*A* and *B*) Computed tomography of the chest with contrast demonstrating enhanced enlarged bronchial vessels (*arrows*) adjacent to the aorta (*A*) and within the pleura (*B*). (*C* and *D*) Low-magnification (4×) view of selected serial hematoxylin and eosin–stained sections of a respiratory lobule surrounded by pleura (Pl) and interlobular septum (Is) that contains a pulmonary vein (PV). Pleura has enlarged bronchial arteries (BA) with a large bronchopulmonary anastomotic vessel (IBA) connecting the BA and pulmonary artery (PA). (*E*) High-magnification (40×) view of respiratory bronchiole (Br) adjacent to a lobular PA with surrounding alveolar spaces containing numerous foamy macrophages (m) filled with cholesterol crystals (c) diagnostic of lipoid pneumonia. (*F*) Three-dimensional reconstruction of a respiratory lobule demonstrating a widely open IBA (*yellow*) connecting a lobular PA (*aqua*) to a pleural BA (*pink*). *Brown*, Pl and Is; *green*, bronchiole; *blue*, pulmonary vein; *red*, endothelium.

A 2-year-old female with systemic juvenile idiopathic arthritis, macrophage activation syndrome, and evolving pulmonary disease developed hypoxemia of unclear etiology. Computed tomography showed lower lobe centrilobular nodularity, ground-glass opacities, and prominent bronchial vessels, notably in the pleura (Figures 1A and 1B). Due to refractory hypoxemia, a lung biopsy was obtained, which revealed extensive lipoid pneumonia (Figures 1C–1E), with a widely open intrapulmonary bronchopulmonary anastomosis (IBA) connecting a lobular pulmonary artery with a pleural bronchial artery. Three-dimensional reconstruction of serial histologic sections was rendered (1), confirming the presence of a recruited IBA (Figure 1F). No autoimmune injury was noted on biopsy. With time, the patient's hypoxemia slowly improved, and she was discharged to home.

In postmortem studies, IBA recruitment has been described in congenital heart disease, in inflammatory lung diseases (including pneumonia), in neonatal lung disorders, and in idiopathic pulmonary hypertension, and it is thought to contribute to right-to-left shunt and hypoxemia (2–4). *In vivo* agitated saline studies have demonstrated that intrapulmonary arteriovenous anastomoses can be recruited in healthy humans (5). Here, we demonstrate a recruited IBA in the distal lung of a living patient with lipoid pneumonia. The role of

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IBAs may include local regulation and redistribution of blood flow in high resistance regions of non-functioning and diseased lung parenchyma.

Some of these results have been previously reported in the form of an abstract (6). ■

Author disclosures are available with the text of this article at www.atsjournals.org.

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