Clinical Image

Exuberant Paraseptal Emphysema Associated With Pulmonary Alveolar Microlithiasis: 12 Years of Evolution

Edson Marchiori a, *, Rosana Souza Rodrigues a, b, Miriam Menna Barreto a

a Universidade Federal do Rio de Janeiro, Rio de Janeiro, Brazil
b Instituto D’Or de Pesquisa e Ensino, Rio de Janeiro, Brazil

A 32-year-old non-smoking man with pulmonary alveolar microlithiasis (PAM) was admitted with cough and dyspnea. Laboratory test results were unremarkable. Chest computed tomography (CT) revealed paraseptal emphysema compromising the entire extent of the subpleural pulmonary parenchyma, and calcifications predominating in the paramediastinal regions (Fig. 1A–C). CT performed 12 years previously showed similar alterations, to a lesser extent (Fig. 1D). Pulmonary biopsy performed 12 years previously showed multiple laminated microliths filling the alveolar spaces compatible with PAM (Fig. 1E). The patient was managed on an outpatient basis.

PAM is a rare autosomal recessive disorder characterized by the intra-alveolar accumulation of spherical calcified concretions (called calciferites or microliths) in the absence of any known calcium metabolism disorder. Most patients with PAM are asymptomatic at the time of diagnosis, and the disease is usually detected incidentally during routine examinations. CT studies of PAM frequently demonstrate the presence of subpleural cysts with diameters of 5–10 mm, which may represent early lung fibrosis. These cysts correspond to the “black pleura” chest-X ray sign. The CT findings of PAM are so characteristic that additional diagnos-
tic investigation is usually unnecessary, especially in patients with other family members with PAM.\textsuperscript{1,2}

**Conflicts of interest**

The authors declare that they have no conflicts of interest to express.

**References**