

Tuberous Sclerosis Complex with Lung Involvement

Arthur Robert Kurzbuch¹ Jean-Yves Fournier¹

¹Department of Service de Neurochirurgie, Hôpital du Valais, Centre Hospitalier du Valais Romand (CHVR), Hôpital de Sion, Sion, Switzerland

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A 31-year-old woman, nonsmoker, known for tuberous sclerosis complex (TSC) underwent uneventful endoscopic resection of a growing subependymal giant cell astrocytoma localized in the left lateral ventricle. Two days after the operation the patient developed dyspnea. Computed tomography of the chest revealed a left-sided pneumothorax (blue asterisk) and diffuse cystic changes throughout the lung parenchyma (blue arrows) consistent with pulmonary lymphangioleiomyomatosis (LAM; ► **Fig. 1**). TSC is an autosomal dominant disorder, resulting from mutations in chromosome 9 (TSC1 encoding hamartin) or chromosome 16 (TSC2 encoding tuberin), that manifests with multisystem involvement: skin, brain, heart, kidneys, and lungs. LAM affects predominantly women and is characterized by proliferation of abnormal smooth-muscle cells in the lung parenchyma that are responsible for cystic changes. Clinically, patients present with dyspnea and pneumothorax. The pneumothorax required draining for 4 days and the patient was discharged home 2 days later without further complications.

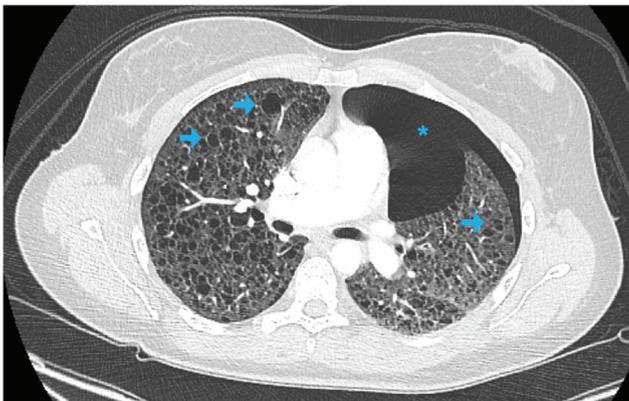


Fig. 1 Axial computed tomography of the chest shows diffuse cystic changes throughout the lung parenchyma consistent with pulmonary lymphangioleiomyomatosis (blue arrows) and a left-sided pneumothorax (blue asterisks).

Address for correspondence Arthur Robert Kurzbuch, MD, Department of Service de Neurochirurgie, Hôpital du Valais, Centre Hospitalier du Valais Romand (CHVR), Hôpital de Sion, Avenue du Grand-Champsec 80, CH-1951 Sion, Switzerland (e-mail: kurzbuch@web.de).

Conflict of Interest

None declared.

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