Internal Medicine
The Japanese Society of Internal Medicine

doi: 10.2169/internalmedicine.5116-20
Intern Med 59: 2801-2802, 2020
http://internmed.jp

【 PICTURES IN CLINICAL MEDICINE 】

Lymphangioleiomyomatosis Associated with Tuberous Sclerosis Complex

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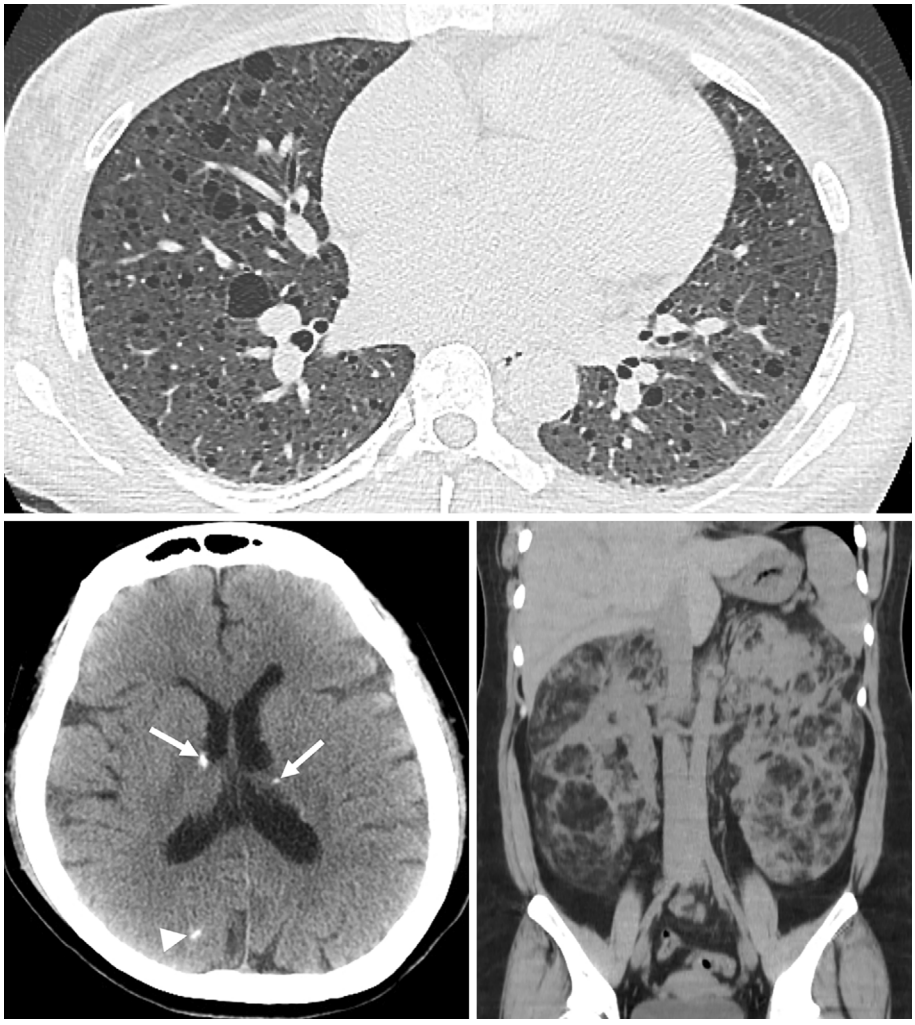
Key words: lymphangioleiomyomatosis, calcified tuber, renal angiomyolipoma, tuberous sclerosis complex, mammalian target of rapamycin

(Intern Med 59: 2801-2802, 2020)
(DOI: 10.2169/internalmedicine.5116-20)



Picture.

A 31-year-old schizophrenic woman presented to the emergency department with asthma-like dyspnea. Laboratory tests revealed mild anemia, and her room air arterial oxygen saturation by pulse oximetry (SpO_2) was 93%. After receiv-

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Received: April 19, 2020; Accepted: June 1, 2020; Advance Publication by J-STAGE: July 14, 2020
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ing bronchodilator drugs, her symptoms improved and the SpO_2 increased to 98%. A subsequently performed computed tomography scan revealed multiple thin walled cysts and increased pulmonary parenchymal attenuation in the bilateral lungs, subependymal (arrows) and cortical (arrowhead) calcification of the brain, and both kidneys were markedly enlarged with normal tissue being replaced by extensive fat-attenuating tissue (Picture). Lymphangioleiomyomatosis, calcified tubers, and renal angiomyolipomas associated with tuberous sclerosis complex (TSC) were diagnosed based on the Japanese TSC diagnostic criteria. Her forced expiratory volume in 1 second was 2.08 L (76.4% predicted) after bronchodilator administration. Transcatheter arterial embolization to prevent the onset of hemorrhaging from angiomyolipomas (1) was not performed in consideration of the risk of renal function impairment, but instead treatment with a mammalian target of rapamycin inhibitor (2) was initiated.

The author states that he has no Conflict of Interest (COI).

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