

Clinical Image

Resolution of Thoracic and Abdominal Lymphangioliomyomas in a Patient With Lymphangioliomyomatosis Treated With Sirolimus[☆]



Resolución de los linfangioleiomiomas torácicos y abdominales en una paciente con linfangioleiomiomatosis tratada con sirolimus

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A 53-year-old woman was diagnosed with lymphangioliomyomatosis (LAM) in 2010, presenting diffuse pulmonary involvement with multiple fine walled cysts, right retrocrural cystic lymphangioliomyomas, and multiple retroperitoneal tumors of

cystic appearance, consistent with abdominal lymphangioliomyomas, the largest being $6.8 \times 3.9 \times 5.1$ in diameter (Fig. 1 A and B). Lung function on diagnosis showed forced vital capacity (FVC) 3300 ml (98%), forced expiratory volume in 1 second (FEV₁) 2370 ml

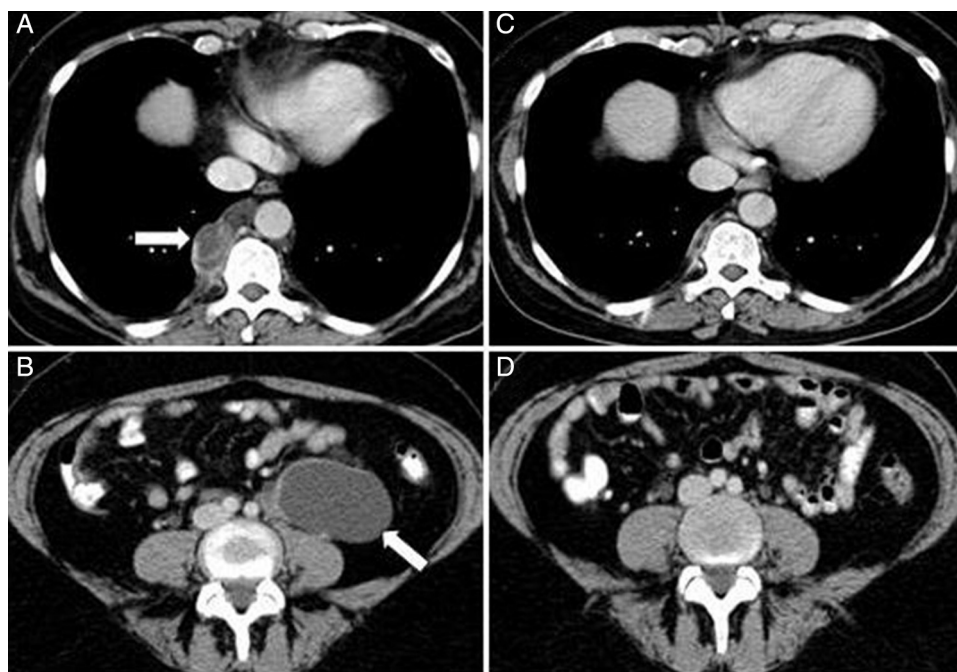


Fig. 1. Chest computed axial tomography (CAT) (A) showing a right paravertebral cystic lesion 3.2×1.9 cm, consistent with retrocrural lymphangioliomyoma. Abdominal CAT (B) showing tumor $6.8 \times 3.9 \times 5.1$ cm of retroperitoneal cystic appearance, located in the left para-aortic region, extending to the pelvis, consistent with abdominal lymphangioliomyoma. Chest-abdominal CAT one year later (C and D) showing practically complete resolution of lesions.

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(82%) and an FEV₁/FVC ratio 69%. Carbon monoxide diffusing capacity (DLCO) was 89%. She reported symptoms of dyspnea on heavy exertion and intermittent episodes of predominantly catamenial chest and abdominal pain. After 2 years of follow-up, lung function had significantly worsened, with FEV₁ 2040 ml (71%) and DLCO 72%, increased basal dyspnea, and catamenial symptoms persisted. In view of her clinical and functional progression, treatment was started with the mammalian target of rapamycin (m-TOR) inhibitor sirolimus at a dose of 2 mg/day.¹ One year after beginning treatment, a follow-up radiological examination showed practically complete resolution of the thoracic and abdominal lymphangiomyomas² (Fig. 1C and D). Clinical improvement and stabilized lung function were observed, with FEV₁ 1980 ml

(70%) and DLCO 82%. This image shows the beneficial effect of sirolimus in reducing the size of the thoracic and abdominal lymphangiomyomas, and in stabilizing lung function in patients with LAM.

References

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