



## Letters to the Editor

### Cystic Lung Disease: The Importance of a Multidisciplinary Approach<sup>☆</sup>



#### *Enfermedad quística del pulmón: la importancia de un enfoque multidisciplinar*

To the Editor,

We read with great interest the well-argued letter to the editor from Herrero et al.,<sup>1</sup> who reported on the association between pulmonary cysts caused by lymphangiomyomatosis (LAM) and abdominal findings in two patients. We would like to highlight some aspects related to these cases.

The main differential diagnoses of cystic disease in LAM include pulmonary Langerhans' cell histiocytosis (PLCH), Birt-Hogg-Dubé syndrome (BHDS), lymphoid interstitial pneumonia (LIP), and *Pneumocystis jiroveci* pneumonia. The distribution of cysts, along with ancillary findings from clinical and imaging studies, may be helpful in distinguishing these entities. PLCH presents with variably sized and often bizarrely shaped cysts. Cysts predominate in the upper and middle lung zones, sparing the costophrenic angles. Nodules in the intervening lung are very suggestive of PLCH, while in the setting of autoimmune disease, particularly Sjögren syndrome, cysts are suggestive of LIP. LIP typically manifests with few thin-walled cysts, predominantly in the lung bases, and parenchymal changes, such as reticulonodular and ground-glass opacities. *P. jiroveci* pneumonia is seen in patients with immunosuppression and typically presents on imaging studies with bilateral ground-glass opacities. Cystic lesions, or pneumatoceles, seen in approximately 30% of cases, are typically transient and occur predominantly in the upper zones. Spontaneous pneumothorax can also occur.<sup>2,3</sup>

Among cystic lung diseases, BHDS is the most difficult to differentiate from LAM, especially when LAM is associated with tuberous sclerosis complex (TSC) with renal and cutaneous involvement. Like BHDS, TSC has a wide clinical spectrum. It is an autosomal-dominant syndrome characterized by widespread hamartomatous lesions. Any organ system can be involved, but the prevalence of manifestations among specific organs differs between children and adults. Patients with TSC usually present angiomyolipomas,

angiofibromas, hypopigmented macules, shagreen patch, and/or periungual fibromas. Cysts in BHDS and LAM are thin-walled, but those associated with LAM are smaller and more circular, homogeneous, and evenly distributed. In BHDS, thin-walled cysts, often larger, are distributed asymmetrically and predominate in the subpleural and paramediastinal regions of the lower lung zones.<sup>2,3</sup>

LAM is typically seen in adult women, as in the two cases described by Herrero et al.<sup>1</sup> In some cases, pulmonary findings alone may not give an accurate diagnosis. However, TSC may be diagnosed without the aid of interventional procedures if associated findings in other organs or systems are carefully evaluated. In view of the abdominal involvement reported by Herrero et al.,<sup>1</sup> these two patients probably presented TSC. Careful histological analysis of the renal mass in the first case was extremely important for diagnosis. In contrast to TSC, in which tumors are usually benign, BHDS carries an increased risk of renal carcinoma. In conclusion, a multidisciplinary approach is required for the diagnosis and assessment of TSC due to the possible involvement of various organs. Assessment and monitoring of patients' relatives is also important, as TSC is a genetic disease.

### References

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