

Images in...

Lymphangiomyomatosis: a radiological diagnosis

Lokesh Shahani

Department of Internal Medicine, Southern Illinois University School of Medicine, Springfield, Illinois, United States

Correspondence to Dr Lokesh Shahani, lokesh83@hotmail.com

DESCRIPTION

Lymphangiomyomatosis (LAM) is a rare disease which predominantly affects young females and is commonly found in association with tuberous sclerosis.¹ The clinical features result from progressive cystic destruction of the lungs and the accumulation of LAM cells within the lungs and axial lymphatics.² The gold standard for the diagnosis of LAM is a tissue biopsy which shows nodular infiltration by abnormal smooth muscle cells, termed LAM cells. Not all patients with LAM require tissue biopsy for a definitive



Figure 1 Computerised axial tomography scan of the chest showing multiple well-defined thin-walled bilateral lung cysts consistent with lymphangiomyomatosis.

diagnosis as the disease has a characteristic computerised axial tomography (CAT) appearance in the majority of cases. The author reports a 34-year-old non-smoking African-American female who presented to the clinic with progressive shortness of breath (Figure 1). CAT scan chest showed multiple well-defined thin-walled bilateral lung cysts which are randomly distributed throughout the lungs with normal intervening lung parenchyma consistent with LAM. There were no infiltrates, worrisome nodules, or pleural effusion. LAM needs to be differentiated from other chronic pulmonary diseases which present with a cystic lung appearance. Langerhans cell histiocytosis which closely mimics LAM is predominantly characterised by nodules and cyst walls of variable thickness and tends to spare the basal parts of the lung near the costophrenic angles.³ Centrilobular emphysema can be differentiated from LAM by absence of well-defined walls and the distribution of the vessels relative to the cystic spaces where the vessels can cross cystic spaces.³

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Johnson SR. Lymphangiomyomatosis. *Eur Respir J* 2006;**27**:1056–65.
2. Johnson SR, Tattersfield AE. Decline in lung function in lymphangiomyomatosis: relation to menopause and progesterone treatment. *Am J Respir Crit Care Med* 1999;**160**:628–33.
3. Seaman DM, Meyer CA, Gilman MD, et al. Diffuse cystic lung disease at high-resolution CT. *AJR Am J Roentgenol* 2011;**196**:1305–11.

This pdf has been created automatically from the final edited text and images.

Copyright 2012 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit

<http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Shahani L. Lymphangiomyomatosis: a radiological diagnosis. *BMJ Case Reports* 2012;10.1136/bcr.11.2011.5192, Published XXX

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow

Keep up to date with all published cases by signing up for an alert (all we need is your email address) <http://casereports.bmj.com/cgi/alerts/etoc>