Pleural Disease in LAM

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The two most common pleural complications encountered among patients with LAM are: 1) Spontaneous pneumothorax, and 2) Chylous pleural effusions

Spontaneous Pneumothorax:

1. Almost two-thirds of women with LAM experience at least one episode of spontaneous pneumothorax in their lifetime
2. Among patients who suffer from a sentinel pneumothorax, there is a greater than 70% chance of recurrence.
3. Pleurodesis (chemical or surgical) reduces the risk of recurrence to approximately 30%.
4. Due to the high recurrence rate, patients with LAM should be offered pleurodesis after their initial episode of pneumothorax rather than waiting for a recurrent event.
5. Although the mode of pleurodesis varies among different centers depending upon individual practices and experiences, we recommend VATS-guided mechanical abrasion as the preferred modality for pleurodesis rather than installation of talc, or other sclerosing agents.
6. Whenever possible, pleurodesis should be handled by clinicians experienced in managing pleural complications of LAM.
7. Patients should be educated about the fact that prior pleurodesis is not a contra-indication to future transplant.
8. All patients should be educated about the signs and symptoms of a spontaneous pneumothorax at the time of initial diagnosis, and advised to seek immediate medical attention if they experience any of these symptoms.
9. The risk of a spontaneous pneumothorax associated with air travel is approximately 1-2 episodes per 100 flights. In general, it is safe for LAM patients to undertake air travel. Patients should be advised to seek medical attention, and not board an air plane if they experience new/worse pleuritic chest pain and/or dyspnea prior to undertaking air travel.
10. Although there is no direct evidence, patients with LAM should be advised against scuba diving due to the potential increased risk of spontaneous pneumothorax related to atmospheric pressure changes.

Chylous Pleural Effusions/Chylothorax:

1. Chylous pleural effusions can be seen in 10-20% of patients with LAM.
2. If safe, chylous effusion should be confirmed with a diagnostic thoracentesis. The presence of chylous effusion in a patient with characteristic cystic changes on HRCT is diagnostic of LAM. Pleural fluid obtained from diagnostic thoracentesis should be investigated for the presence of LAM cells. Although not necessary, the demonstration of LAM cells on pleural fluid cytology further strengthens the diagnosis of LAM.
3. Sirolimus is effective in managing chylous effusion, and should be the first line treatment option for patients with chylothorax, as compared to invasive treatment modalities.

4. We recommend that patients undergo dedicated lymphatic imaging such as heavy T2-weighted MRI imaging to study the lymphatic anatomy prior to pursuing invasive options.
5. Treatment with sirolimus may take a few months (up to 6-12 months or longer) to be effective at managing chylous effusions. Since chyle does not typically produce pleural inflammation/fibrothorax, it may be prudent to wait for sirolimus to take peak effect prior to considering invasive treatment options.
6. We recommend that invasive management such as thoracic duct embolization be performed by clinicians who have expertise in managing lymphatic complications in patients with LAM.

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