Transcript Details

This is a transcript of a continuing medical education (CME) activity. Additional media formats for the activity and full activity details (including sponsor and supporter, disclosures, and instructions for claiming credit) are available by visiting:
https://reachmd.com/programs/cme/ph-ild-diagnosis-and-management/16511/

Time needed to complete: 4h 49m

ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

PH-ILD: Diagnosis and Management

Announcer:
Welcome to CME on ReachMD. This episode is part of our MinuteCE curriculum.

Prior to beginning the activity, please be sure to review the faculty and commercial support disclosure statements as well as the learning objectives.

Dr. Schacht:
So, the next topic we're going to be talking about is a focus on CTD, PAH, and ILD overlap. My name is Sydney Schacht. I'm from Indiana University. I'm a Pulmonary Critical Care Fellow.

So we're going to start with a case here. So we have a 65-year-old woman. She has a history of a cough, recurrent pneumonia, hypertension, Hashimoto thyroiditis, she had a thyroidectomy a couple years ago, bronchiectasis, and she comes to the clinic for evaluation of dyspnea on exertion. So a little bit of history; 4 years ago, she had a cough after her thyroidectomy 2 years prior, so 2 years later, she was hospitalized with pneumonia one time. She required mechanical ventilation. And then in the 6 months leading up, she's had progressive functional decline with dyspnea on exertion. She used to walk 3 miles a day, now she can only really perform ADLs and has to rest afterwards. She has exertional chest pain, no syncope, she does have some edema and weight gain of unknown amount. And her symptoms are definitely worse during the day and with activity. Her cough is not productive. She has GERD; it's somewhat better with a PPI, and bronchodilators really don't seem to help. Her social history is unremarkable.

So on exam, I've only just highlighted the things that were most important so you can note that her tachycardia is low grade. She has scattered telangiectasias on her forehead. She has JVD. She has an S3. And on pulmonary exam, she has basilar crackles. You can see on her skin exam she has some skin tightening and periungual erythema.

So secure some initial diagnostics from early 2021. You can see her CT on the right there. Her PFTs, she had a normal ratio her FVC was 2.58, which was 80% of predicted, her TLC was 77% of predicted, and her DLCO was the most notable, decreased at 8.4 which is 40% of predicted. You can see on the two cuts that are representative that she has some traction bronchiectasis, most notably in the lower lobe.

Again, I've highlighted just the normal or abnormal laboratory data. You can see that her ANA is elevated 1:160. Her anti-centromere antibody was negative, but her SCL 70 antibody was positive at 51. And then we use both NT-pro and BNP depending on the patient, and both of them are elevated to 2797 and 405, respectively. Everything else was unremarkable in terms of her workup.

Other diagnostics, so she underwent home O2 evaluation, okay on room air with rest but desat with exertion, so she was prescribed 1L of oxygen with exertion only. Her VQ scan was negative for a PE. She did have a bronchoscopy during one of her admissions, which unfortunately was not sent for all of our cell counts as usual. But it did show that she had negative microbiology. She had mild sleep apnea and was prescribed a CPAP. And on her 6-minute walk test, by the end of 2021, she could only walk 244 meters on 3L of exertion.

So here's some representative cuts of her echos. You can see that she's dilated, most notably on the right side RA and RV dilatation. She has some septal flattening and both systole and diastole, indicative of pressure and volume overload.

Here's her right heart catheterization around the same time. You can see that all of her pressures are elevated and except for her wedge which is low, so her mean PA was 63, her PVR was almost 15 Woods units. She did not have a response to nitric oxide and her indices, by both thermodilution and Fick, were on the lower end.

So she was started on triple therapy including subcutaneous treprostinil. She was given a diuretic regimen with some improvement in her waist and lower extremity edema. She eventually was diagnosed with scleroderma by rheumatology around the same time and started on mycophenolate and then referred to pulmonary rehab. And she's actually doing really well.

So since starting her therapies and her diuretics, she's improved symptomatically across the board. She went from class III down to a class II in terms of her functional class. And even though she's completed pulmonary rehab many times over, she continues to go and she finds it incredibly beneficial. Her high-risk features on presentation are now considered low risk.

She's had a repeat right heart cath back in December of 2022. All of her numbers have improved, her mean PA went down to 41 and her wedge is still low at 9. Her echo also similarly shows some improvement, but she has developed some stage 2 diastolic dysfunction. Her PFTs are also improved. The only thing that really unfortunately has changed for her is that she now needs 1L of oxygen with rest and 6L with exertion, but that doesn't seem to faze her since she's quite active these days. Similarly, her 6-minute walk test is also improved from 244 to 444 meters.

Alright, and with that, we'll move on to our next speaker.

Announcer:
You have been listening to CME on ReachMD. This activity is jointly provided by Global Learning Collaborative (GLC) and TotalCME, LLC. and is part of our MinuteCE curriculum.

To receive your free CME credit, or to download this activity, go to ReachMD.com/CME. Thank you for listening.