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Case Study: CTD-PAH

Announcer:
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Dr. Cao:
So today I'm presenting a case of a 61-year-old female with likely Sjogren's, CTD-ILD who presented for evaluation for pulmonary hypertension after an echo showing an estimated PASP of 86. A little bit regarding her history, her breathing problems started around age 47, for which she got a lung biopsy showing interstitial pneumonitis, requiring several days of mechanical ventilation, was on prednisone for 2 years. About 6 months prior to presentation, she had an ECG done during an annual physical, where the doctor noted a glitch and basically was referred to cardiology, where she got an echo showing RV pressure overload and this estimated PASP of 86. Two months prior to presentation, she did faint in the setting of feeling dizzy when getting up to go to the bathroom. Her functional capacity at baseline though, she said was unlimited when walking on flat ground, although it did have rare chest pressure and heaviness with exertion. With regards to her rheumatologic history, she has a history of Raynaud's since age 49, history of joint pains and with rash, difficulty swallowing for about 5 years, but she underwent esophageal dilation and ultimately was diagnosed with likely Sjogren's, given her sicca syndrome, positive ANA, and positive SSA. And social history, she smoked for 20 years, 3 cigarettes a day, and quit around age 40.

So looking at her CT chest, we can see that she had bilateral lower lobe greater than upper low course ground-glass opacities, peribronchial reticulations with increased traction bronchiectasis, with some limited subpleural sparing. Overall, the constellation of findings was consistent with NSIP pattern.

Her pulmonary function tests shown here does show moderate restriction with TLC 60% predicted and severe diffusion impairment with a DLCO that was 33% predicted. She was able to walk about 351 meters on her 6-minute walk test.

Did want to show her initial ECG which prompted her pulmonary hypertension workup, and here we can see she had an incomplete right bundle. And then on her repeat echo, my videos aren't going to play, but she basically had a moderately enlarged right ventricular size and moderately reduced right ventricular systolic function. The RV to LV ratio is 1.25, a TAPSE of 1.3. And she also had a mild to moderate tricuspid valve regurgitation with a regurgitant velocity of 4.69, which basically estimate her RVSP to be around 91, which was similar to her initial echo showing the estimated PASP of 86.

So given all these results, she proceeded to a right heart cath, which was notable for a PA pressure of 96/39 with a mean of 58, a wedge of 15. Thermo cardiac output was 1.63, which yielded a index of 1.01. Her PVR was severely elevated at 26. Overall, this was consistent with severe precapillary pulmonary hypertension, and there was no evidence of acute vasoreactivity to iNO.

So I think the suspicion for pulmonary hypertension in this case was fairly high. And it was a pretty clear decision that she needed a right heart cath. However, that decision, I think, is not always so clear cut. So I did want to mention here, the DETECT algorithm which is one of the first evidence-based algorithms for screening patients for PAH mostly in systemic sclerosis. It's a tool that's been used to

identify patients with PAH in asymptomatic stages. So it uses a combination of clinical variables, pulmonary function tests, immunologic, biologic, electrocardiographic, as well as echocardiographic parameters to decide whether or not to proceed with echo, and then ultimately whether or not to proceed with right heart cath.

Announcer:
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