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Case Study: A 62-Year-Old Male With CTEPH

Announcer:
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Dr. Hanna:
Thank you guys for having me. And it's a pleasure to present this case of a 62-year old-gentleman.

So initially, you know, he came to us as a 62-year-old gentleman but initially presented as a 60-year-old gentleman with no past medical history, no follow-up with any doctors, and actually had a provoked PE after falling and slipping on ice, and then shortly after driving from Michigan to Texas. He presented in Texas. Here you can see his CTA kind of showing pulmonary emboli, and significant blood work included in elevated troponin and BNP as well.

So in the hospital, he was started on low molecular weight heparin for anticoagulation and then transitioned to apixaban. Again, because he had never really been assessed by a physician, he was diagnosed with a myriad of new medical problems and discharged on a bunch of other medications. He was still dyspneic 3 months after presenting, and in an outpatient follow-up, outpatient PFTs were overall unremarkable, but still quite short of breath. And a few months later, he actually presented to the emergency room again for expressive aphasia. This lasted around 10 minutes. He was adamant that he had not missed any of his anticoagulation. And by the time he presented to the emergency room, he was back to his baseline. In the ER, he had negative CT/CTA. But he was admitted and underwent an MRI of the head and that did confirm an acute stroke in the distribution of the left MCA. Because he was admitted to the hospital and they wanted to rule out other potential etiologies of stroke, he did undergo an echocardiogram. And I include this again, because this is a case of CTEPH, and it showed persistent RV enlargement and dilation with a reduced systolic function. And he had McConnell's sign. Again, his pulmonary embolism was around 5 months back.

So his statin was up-titrated, he was started on aspirin in addition to his apixaban for a CVA, and again, for persistent shortness of breath and the fact that he had PEs, he was referred to pulmonology.

As part of the workup, even though this was in the provoked setting, he did undergo a hypercoagulable workup with a repeat echocardiogram. His hypercoagulable workup was positive for prothrombin gene A, IgG and IgM for cardiolipin, and he also tested positive for ANA and anti-double-stranded DNA antibodies.

His echo now almost 8 months after additional persistent RV dilation, and again, more importantly, he was persistently symptomatic. Because he was persistently symptomatic, he was evaluated for CTEPH with a VQ and a repeat CTA of the lung. The VQ was positive for multiple wedge-shaped perfusion defects. And a CTA did show an improvement in his clog burden; however, he did have residual PE within his left PA and a few proximal bilateral subsegmental branches.

So he was referred to University of Michigan at the Pulmonary Hypertension Clinic, again for his persistent dyspnea and the positive scans. He was still quite below his baseline. We classified him as WHO functional class II. He underwent 6-minute walk test, a right

heart cath, and he walked 378 meters, so kind of in that low-intermediate classification, is 63% of the average. And he didn't really desaturate. His right heart cath did show, and again I'll just focus on his wedge, his mean PA, wedge below 15, and mean PA above 20, and again, PVR well above 2 or 3.

With the positive VQ scan, he underwent pulmonary angiography. This is the right lung progression, the RAO and LAO, and you can actually see, if I go back, a relative perfusion defect in the lower zones. You can kind of see contrast hitting the periphery but not so much in the lower zones. Again, just not in too much detail. And the left lung, same thing. And in addition to already being on apixaban, he was transitioned to warfarin, and he was started on riociguat.

So seen in the BPA clinic, and after discussion, he remained symptomatic, and he was evaluated by our BPA team, multidisciplinary CTEPH team, which includes a interventional cardiologist, cardiologist with a specialization in pulmonary hypertension, interventional radiology, and CT surgery. And depending on the location of his burden, he was deemed not to be a candidate for endarterectomy because it was not proximal disease, and elected to undergo BPA therapy, which we'll review that in a few minutes.

Next up is Dr. Ruben Mylvaganam. He's going to talk to us about the epidemiology and clinical trends in CTEPH and CTED.

Announcer:

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