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

Pulmonary Hypertension in Rheumatic Diseases - Part 2

Announcer:
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Dr. Kafaja:
Again, I discussed earlier about the differences between the diffuse and limited. And this just basically goes to show that there are differences among our patients with the diffuse presentations versus limited presentation and what sort of antibodies they might have.

So for the patients that have limited presentation, limited systemic sclerosis, again, sparing the upper arm, sparing the thigh, sparing the torso, you might see patients having anti-centromere antibodies, you might see them having PM/SEL antibodies, Th/To, or U1-RNP.

In the diffuse patients, you're more likely to encounter patients with SCL-70, RNA polymerase-III antibodies, which in those patients are at high risk for developing scleroderma, renal crisis, as well as it's been associated with the development of malignancies pre, during, or post their diagnosis as well as you U3-RNP.

But interestingly, there was this particular study, they've actually shown that there are specific antibodies that they look for, were saying similar to what our previous colleagues in their previous session were discussing. There are some antibodies that do clue you in. And this is the anti-centromere antibody in the setting of scleroderma, for instance, U3-RNP, Th/To, and anti-phospholipid. And lupus patients in this particular study, they found them to have U1-RNP, anti-phospholipid, as well as SSA/SSB.

And what I wanted to point out, this is the most recent study actually was published in 2023, I wanted to highlight this one even though I skipped it initially. And this is an anti-centromere, is a subset of centromere antibody, anti-centromere p4.2 antibody, where they found them to be associated with a decreased diffuse capacity, regardless of whether patients had ILD or not. And this may be one of the kind of the up and coming antibodies that we should be looking at, or maybe that would give us a clue as to whether these patients may be more likely to have pulmonary hypertension down the line.

And why we focused, I know that Dr. Saggar had discussed this earlier, and we're focusing on pulmonary hypertension and interstitial lung disease, and even though these patients - what is the significance of these patients developing this? In scleroderma specifically over the years, we have noticed that there has been a shift in the patient's presentation, where in the past, patients to use to die. You know, we talked about morbidity versus mortality, right? And in the past, patients used to develop a lot more renal crisis, for instance. You see it here. And then that seems to have improved over the years by us recognizing, a rheumatologist, as well as specialists all throughout medicine, recognizing this entity early on, so that we're treating patients and then also paying attention to not use, for instance, steroids so freely, especially in patients with systemic sclerosis, when that is - when that diagnosis is in question. But what we're also seeing over the years is that pulmonary hypertension patients as well as ILD are, this is the cause - these are the cause of death among our patients. And this is why we are here today discussing this.

Along the same line, we also noted that patients with pulmonary hypertension over the years, even though this is an old study, but this

seems to be continuing on, is that patients with pulmonary hypertension versus those with lung involvement only without pulmonary hypertension, seem to have the worst outcome.

Now, this brings us back to is there an interplay between ILD and pulmonary hypertension? As Dr. Saggar was - you were alluding to that one before and whether we should be paying attention more to those subset of patients. And I think based on this, I would argue yes, and based on all the data that was presented earlier. And now we have these patients in this particular study, they found that patients with when they focused on SSc ILD patient, they noticed that they - these subsets of patients are the ones that develop severe pulmonary hypertension, were sixfold - it had a sixfold increase a hazard for mortality. And then those with moderate had about twofold increase. In looking at doing a univariate and multivariate analysis, when they looked subsequently at patients with systemic sclerosis, pulmonary hypertension, and they looked at the subset of patients that had severe versus moderate interstitial lung disease, again, severe ILD was associated with a threefold increase hazard for mortality in these patients. So there is an interplay between the two. And we should definitely, you know - it has implications for our patients for the future and their outcomes. So, I think the sooner we start them on therapy, is of utmost importance.

Now, what are some of the predictors then to help us guide the patients to get to you? Some of the things that we pay attention to is the slow decline in DLCO. Of course, there's been reported the FVC to DLCO ratio of more than 1.6, as well as now with the changing of the of the guidelines, the question is how is that changing our referrals to our colleagues, our pulmonary colleagues, our pulmonary hypertension colleagues, as well? I did discussed anti-centromere and I hope that I honed down to, you know, centromere is associated usually with the more vascular component. So it's not surprising that it is - that would be included too in our kind of our highly likelihood to more - to send patients to you, patients with limited disease, as well as extensive telangiectasias, those are broken blood vessels or dilated blood vessels that are apparent sometimes in their mouth, on their face, hands, chest. So this is all indicative of vascular subtype, if you will.

Now, and over time, though, what has been noted is that a patient - when they looked at this patient - these patients, this is from seen Jenny Steen. And this is an old study in 2005, but what I discussed with you earlier is that patients are - it's going to take a while for our patients to get to you or for them to develop pulmonary hypertension. However, it is important to capture them early, rather than wait. And though it takes about somewhere between 10 to 15 years for them to be diagnosed with pulmonary hypertension.

I will go into this one, I think I've already gone into this. But so how do we screen patients? And screening, really is it useful? Should we be doing this? And while this is old news to a lot of you, in this particular study, they looked at patients who were just routinely followed in practice versus those that were screened on regular basis and looked at their survival. And in those who were screened, there was a marked difference in survival versus routine practice. So you have 17% versus 64% are surviving, favoring of course the patients that are routinely screened.

Now we'll discuss the NT-proBNP. It's old news, but it is - this basically goes - it is important to obtain it. We do use it, I don't know how much, you know, sometimes the NT-proBNP is hard to get for us and we resort to the BNP. It is a send-out, but it is a useful tool. And based on the guidelines, it seems that it's even more useful, perhaps even with the - as far as the changing of the of the guidelines, perhaps, you know, the numbers may mean something different nowadays versus before. But regardless, and I think it may mean something different, and I'll bring this up in our discussion at how much of - how much do we do this in clinical practice when our patients get to you.

Now, we all know this, that echocardiograms are not a useful tool in predicting pulmonary hypertension, but we do - this is our first kind of go-to, a way or to evaluate patients. And in this particular study, they did notice that changes in RVSP have about 2 mmHg per year, were actually supportive of things to come. Or, you know, those are the category of patients that perhaps we should be looking into. This is an old study, but I wanted to then move on to this and then just bring up this would be my last slide, I believe, really to bring up the DETECT algorithm. And how much of it do we use nowadays? I know that Dr. Cao had a presented this in her discussion of our case, but the patient that we had, had Sjogren's. And how much of that do we actually use this? This was particularly done in patients with scleroderma, and to evaluate patients with scleroderma where it's a stepwise fashion to kind of help guide referral to right heart cath in these patients. And the question I'll leave you with is how much of that has changed? And how much of that is - should be changing with the new guidelines? But I think the main thing moving forward, I think, I hope, I have shown you that pulmonary hypertension can present in a number of rheumatologic conditions, not just scleroderma. It's important to keep in mind, you know, sometimes you do see patients before we see them. And I get a lot of patients, for instance, sent from Rajan to me, with the question: does this patient have, you know, this patient came to me with pulmonary - severe pulmonary hypertension, but I do notice these hyperpigmentations. I do notice the skin thickening. So, does this patient have scleroderma? Does this patient have some other autoimmune disease that has been missed over the years? Cardio - capillaries are important to look at. And you know, we're starting to actually look at them much more religiously in clinic. So I think it's important, it's a very useful tool, it's very, very fast, and an easy way to just look at the capillaries. It just takes some practice. And I learned it. I have a like a hand tool actually that I purchased on Amazon, $20. And it gives you 200x

magnification, fantastic. Those pictures came from that tool. And I can - I put them in patient's charts nowadays. So there's documentation there's documentation of what happens over time with these patients, and that can help you, at least it helps me to kind of figure out what, you know, when to send my patients, when to start - have that red flag come up, if I start definitely to see these giant capillaries or more of these tortuosities going on. And as far as when it comes to evaluating patients with interstitial lung disease, I hope I honed in on the importance of biomarkers and some of these antibodies that we, you know - I've alluded to before of some new ones that are coming up and hopefully that will help also drive our patients into your clinics much sooner. And of course, you know, early detection is important. So patients that were followed just haphazardly are not doing well. We need to continuously just hone in on getting those PFTs, getting the echoes, looking at RVSP trend, and then of course TR jets, and so on. And I'll leave it out there as far as whether we use the DETECT and how useful it is, not only scleroderma, for scleroderma patients, but is it really applicable anywhere else?

Thank you for your attention.

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