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Pulmonary Hypertension in Rheumatic Diseases - Part 1

Announcer:
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Dr. Kafaja:
Hi, I'm Suzanne Kafaja, a Rheumatologist, and I would like to thank Dr. Channick and Dr. Saggar for inviting me for the Organizing Committee, and for all of you for being here.

What I wanted to discuss today is really an introduction to pulmonary hypertension in the setting of rheumatic diseases; and discuss how prevalent that is in connective tissue disease spectrum, as well as focusing on systemic sclerosis specifically, which is my area of expertise and passion. We'll discuss briefly the usefulness of autoantibodies. We have - I think the previous panel had discussed briefly the usefulness of some antibodies that you get and when to get them. And I know that Dr. Saggar and other colleagues have discussed this as well as far as the ILD PAH interplay, so I'll touch upon that as well.

So as you all know, when discussing pulmonary hypertension, the focus typically is to jump on to systemic sclerosis. But I wanted to bring out to your attention that when looking at all the different groups of pulmonary hypertension, there are various rheumatologic conditions that can present with that, including lupus, systemic sclerosis, of course, as well as Sjogren's, sarcoidosis, and others. So to keep in mind, you know, this is kind of a - it goes to show you that whether it's Group 1, 2, 3, or 4, or 5 for that matter, the pulmonary hypertension presentation can present in a number of rheumatologic conditions. So, we see here that for Group 1, for instance, mainly it's the systemic sclerosis patients that present that way. Group 2, the focus is sometimes mostly on rheumatoid arthritis, but also systemic sclerosis as well. Group 3, really an umbrella of diseases that can present with that, including dermatomyositis, polymyositis, which is something that I have not discussed earlier, as well as systemic sclerosis and sarcoidosis. These are things that were brought up by Dr. Saggar earlier and the previous group, as well as, you know, keep in mind that we have the thromboembolic conditions which are related to Group 4 and patients with lupus or just antiphospholipid syndrome. And keep in mind that even a subset of other conditions including scleroderma patients, can have antiphospholipid antibodies that are positive and should be looked into. About 10% of patients with scleroderma can have it, up to 10%. So make sure to include that in your - in the sort of antibodies and that you look for in this subset of patients. So the bottom line is that any pulmonary hypertension group may be detected in patients with CTD. So keep that in mind when evaluating your patients, not just Group 3, not just Group 1; so all of it.

And then when focusing now on, you know, and looking at the scleroderma specifically and why we focused on scleroderma specifically, this is a study basically that looked at the French registry as well as the Reveal registry which is a U.S. group. The combination of this is about 3,000 patients where the sum of this basically goes to show that systemic sclerosis patients associated pulmonary hypertension account for about 75% of all the cases, but keep in mind that yes, other connective tissue diseases are involved. And why do we focus on these groups? You know, and what age should we focus on them? Really, systemic sclerosis patients are predominantly women; and this is what's been noted here is that the majority of the presentation occurs in women, anywhere between a ratio between 1.9 to 4 depending on the registry that you're looking at. And age at presentation is about 50, at least based on the registry.

Now focusing on systemic sclerosis. It's not – it's an umbrella of conditions that patients can have. So while our patients can have a number of organs that are in evolved, so their presentation can vary. So some of our patients have skin thickening, some don't. Some have lung involvement and lung fibrosis, some don't. Some have GI involvement where - and this actually, you know, I don't think we discussed this earlier, but it does take into – you know, we need to take it into account and it should be taken into account. It is taken into account when patients are sent for a transplant, for instance, but it can also impact interstitial lung disease and the progression of it. So we need to pay attention to and keep in mind that patients for instance, with systemic sclerosis as well as myositis patients, can have esophageal dysmotility. And so they can have reflux, they can have micro aspirations that could actually make it look like their interstitial lung disease, for instance, is getting worse, while it could be contributing to some of their dyspnea symptoms that could be complicating the picture for us. Patients with scleroderma also can have renal involvement, renal crisis. So they can present with proteinuria, pulmonary - with hypertension. So that can also lead to – with their proteinuria, if they're having renal crisis, their creatinine rises, and sometimes that can foggy up the picture for us. And we need to pay attention that, you know, perhaps when they present with dyspnea on exertion, it's not necessarily - and especially when it's sudden, it may be related to any number of these symptoms. Patients can have a dryness in their eyes, mouth, and secondary Sjogren's as well. Musculoskeletal involvement, as far as our patients go, they can have muscle weakness, they can have myopathy, they can have overlapping myositis. So that all plays into perhaps their ability to expand the lungs, they expand their chest wall, and their overall fatigue and dyspnea. So not everything is surely a presentation - a straightforward presentation or something that we're going to be able to make a determination based upon that.

And lastly, I want to point out the Raynaud's, and I'll talk about this in detail in a little bit, where this is a big focus, and I would like to hopefully, with the some of the points that I'm going to point out to you, and some of the take-home messages for you will be to pay attention to some of these patients that are presenting with more of a vascular presentation, because those are the patients that we worry about as far as pulmonary hypertension.

So given what I've just told you, patients with systemic sclerosis can have a multitude of causes for their dyspnea or fatigue or chest pain presentation or palpitations. And when we look at the causes, of course, we see that it could be due to restrictive lung disease or myopathy as discussed earlier, it could be too that their heart failure, for instance, and LV diastolic dysfunction, as well as anemia due to GAVE, for instance, gastric ectasia, so they could be losing blood, becoming dyspneic. Which also again, I think goes to show that this all complicates the picture of when you evaluate patients with autoimmune diseases.

So, and why we pay attention to pulmonary hypertension is because this pulmonary hypertension is actually one of the major classification criteria for a systemic sclerosis; it is something that we don't take lightly. And although it takes a while for patients to present, it might take years for them to present. But I'll go into what type of patients we need to evaluate more closely.

So these are some of the differences. You are probably all familiar with this and the difference between patients with systemic limited disease versus diffuse. So patients with diffuse disease are more likely to develop as time goes on, years down from their presentation. They tend to have more pulmonary fibrosis, less so of pulmonary hypertension, less so of scleroderma, renal crisis, or cardiac involvement. If you look at the patients with limited disease, those are the patients with more vascular components of their disease, they're more likely to develop less so than the others, they develop pulmonary hypertension or pulmonary fibrosis but more so of them develop pulmonary hypertension compared to the ones with the diffuse disease. The scleroderma and renal crisis is very, very minimal compared to others. And as far as the cardiac disease, it's about equal.

So some of the things that we take into account when evaluating patients with pulmonary hypertension is what is their age. I just did discuss with you that patients who are usually perimenopausal, around year 50, is typically women are the ones that we focus on historically, new onset digital ulcers. So this is, again, more the vascular patients that are presenting with - that are at risk for developing pulmonary hypertension, those that are presenting with limited systemic sclerosis. So skin that is basically sparing their upper extremities, sparing their thighs, and their abdomen, those are the patients with limited scleroderma. Dilated nailfold capillaries that we'll be discussing soon.

So this is a patient with Raynaud's that I have. And you see the discoloration, the purple discoloration of her hands. And the classical definition of Raynaud's is really episodic, symmetrical, vasospastic disorder, that involving a triphasic color change in patients. So they go from white, red, purple, sometimes white, red, blue, but classically, it's a triple change in the color. And there are variations. And I bring this up, I know this is maybe kind of a too date – too detailed for a majority of the group here who is probably a pulmonologist. But I wanted to really bring this out to your attention because capillaroscopy really plays a role in our assessment of how likely for patients, you know, I know Dr. Channick had discussed the new guidelines and it would be nice for a rheumatologic kind of input into this, especially for the vascular component of patients and where we stand. Because typically, for patients that don't have nonspecific abnormalities in non-scleroderma patients, they - you will tend to see that their capillaries - so typically, when we look at the nailfold capillaries using multiple gadgets, sometimes just an ophthalmoscope that we have in the office, we looked at as to how well defined,

and it's almost like all the vasculature needs to look like picket fence, the blood vessels in nailfold should look like picket fence. And whenever we see decrease in the density as well as decrease in change in the pathology, for instance, you might get dilated blood vessels, and I'll go into this a little bit further, that may be a clue as to where the patient is headed; and whether they have scleroderma or they don't have scleroderma, that may be a clue that they're headed towards a more a vascular presentation down the line. In patients with scleroderma, for instance, they're an early on, you don't have any decrease in the density. But later on in patients with active disease, you might start to see some giant capillaries presentation and then you might see a little bit of dropouts later on. So then these blood vessels start to disappear on you.

In this particular study, for instance, they noted that a higher number of patients with systemic sclerosis who develop pulmonary hypertension had more severe patterns of nailfold capillaries where they had decreased density, increased main loop capillary width, so they start to develop these giant capillaries and they get these neoangiogenesis where you get new capillary formation.

And these are some examples, for instance, that I've taken in my office off of patients that I saw, where you have - this is the picket fence that I was talking about. You see how well organized the capillaries are. And in some patients, you start to see early on these capillary leaks. Later on, and these are some of the patients that I would start to get worried about into developing pulmonary hypertension or these are more vascular patients, more vascular presentation. This is a giant capillary here and you start to see the dropout, so the capillaries are not as dense, they are becoming far in between, and you get these giant capillaries and this ballooned-up capillaries as well. So those are patients that I would argue that I would worry about them into developing more vascular presentation like pulmonary hypertension down the line.

So these are just different patient's presentations where you see even undifferentiated connective tissue disease patient presenting with hemorrhage. Another one with lupus. You see the - just the variations in the capillaries and sometimes you can, you know, those are the type of patients that you might start to kind of pay more attention to down the line that they are at high risk, perhaps, of developing this.

Again, along the same line, this is very later on and in very severe cases you might develop - you might have patients that develop digital ulcerations and ischemia, dry gangrene, and they might develop an amputation. So those are the patients you need to worry about.

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