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The Sarcoidosis-PAH Patient – A Rare and Challenging Situation

Dr. McLaughlin:
Hello. Today we're discussing sarcoidosis as one of those causes of pulmonary hypertension which is really underrepresented in our clinical trials. I'm Vallerie McLaughlin from the University of Michigan, and I'm joined today by my friends and colleagues, Dr. Sudarshan Rajagopal from Duke, Ioana Preston from Tufts, and Rich Krasuski from Duke as well. You guys, thanks so much for joining me.

Ioana is our only pulmonologist here, you probably know the most about sarcoid of all of us. Do you want to tell us a little bit about the histology and the basis of sarcoidosis?

Dr. Preston:
Vall, sarcoid is a fascinating disease and it can affect the lungs in 90% of patients with sarcoidosis. And within the lungs, it can affect the interstitium, it can affect the bronchial tree, and the pulmonary vasculature. There is a subpopulation of sarcoid patients who develop pulmonary vascular disease that is very similar to our Group 1 PAH patients. And those are the patients in whom we can focus on treating with targeted therapy.

But if you look at the histology, if you ask me, it's very similar to scleroderma in the sense that the granulomas of sarcoid involve the pre-capillaries, the capillaries, but also the venules of the pulmonary vasculature. So it's a much more extensive vascular involvement.

Dr. McLaughlin:
So to me, that sounds like it can cause pulmonary hypertension in a lot of different ways. Do you want to review that for us, Rich?

Dr. Krasuski:
Sure Vall. So obviously it's a complex disorder. It's also a big mimicker of a whole variety of different disorders. It can not only cause pulmonary hypertension, but it can cause systemic disease as well. I mean, sarcoid is a very complex disorder, but as far as PH goes Vall, it can affect almost any type.

So you can have Group 1 disease, of course, which is what we're mainly focusing on today because that responds to advanced medical therapies. But similarly, you can get sarcoid-related heart failure, that's not unusual. You can get a cardiomyopathy from sarcoid and that's important to distinguish, particularly, before you start advanced medical therapies for PH cause it could be a mixed picture. You can get Group 3 disease, which Ioana, very appropriately talked about. I've certainly seen my even share of patients with thromboembolic phenomenon related to sarcoid. And finally, the Group 5, and the reason it's been grouped into Group 5, because the mechanism is still not well understood.

So really you could place it into any of those five groups but ideally, you really want to look for a Group 1 component before you start treating with these therapies.

Dr. McLaughlin:
Right, so again, you're right. I mean it's in Group 5 primarily because it is multifactorial. And so it can cause any of those types of pulmonary hypertension, I think.

Sudar, what's your approach? Like, let's say you get referred a patient with known sarcoid. Again, as Ioana said, they almost all have

pulmonary involvement, this has happened to all of us, right? They're followed by pulmonary, they feel like their shortness of breath is worse, but the pulmonologist says the sarcoid is stable. What's your approach to that patient?

Dr. Rajagopal:
So, in general, the real focus is on diagnosis here and that diagnostic approach. The right heart cath is critical here because you really want to see high PVR that you'd expect to see in Group 1 pulmonary hypertension. Ideally, also, you don't want to see a high wedge which would suggest that you have sarcoid involving the heart. But you could also rule out using PET or cardiac MRI, looking at the PFTs and trying to assess what degree of parenchymal lung disease they have. Although, I think we've all had experiences in treating even patients with significant parenchymal lung disease with sarcoid. If they have a high PVR, they could still even have a response to PAH-specific therapies.

And then it's really about starting low and going slow in terms of treating these patients, being careful. Once you've made that decision, that they have a significant precapillary component to their disease, you can start therapy and just be careful in ensuring that they're not developing worsening hypoxia and that they're responding well. And there are some observational studies from Hap Farber's Group, from the University of Chicago and from Duke that have shown that patients can tolerate these therapies well and even have some improvement in their hemodynamics and INT-proBNP.

Dr. McLaughlin:
Yeah, that's great.

Rich, anything we should be aware of when we do cath, any hemodynamic pearls you want to highlight for these patients?

Dr. Krasuski:
Yeah, I think the same pearls I often talk about for everybody. And that is, sometimes we get to these patients late in the day and we're doing a hemodynamic assessment and we probably don't assess the LV diastolic component as well as we should. So, giving them a volume challenge is not an unreasonable thing if there's any question about their myocardium not being normal. And I oftentimes use the vasodilator challenge the same way because in somebody who is low volume state you can actually identify this by increasing pulmonary blood flow and their wedge goes up or LVEDP goes up, you're recognizing there's other disease that's present.

Dr. McLaughlin:
Right. So good pearls.

So Ioana, said, you know, there's a little bit of data, some observational data, so how do you take that and approach it in your clinical practice? Do you still do risk assessment the way we do in Group 1 patients? Do you try to stick with the therapies that maybe have been included with registries? Just on a practical basis, what do you do every day with these folks?

Dr. Preston:
It's a good question, Vall. So, for the screening purpose for these patients, a decreasing six-mile walk test coupled with a new or worsening desaturation during exercise is a trigger for me to think about pulmonary vascular disease in sarcoid. And then we go for a careful right heart catheterization assessment.

But once we diagnose pulmonary vascular disease in these patients, we treat very similarly with Group 1. And be cognizant that any underlying interstitial lung disease that they may have may contribute to the side effects of our treatments. But, all in all, in my experience, is that they do respond well to PAH therapies.

Dr. McLaughlin:
I don't know about you guys, I kind of bucket these patients, right? I'm not a pulmonologist, I don't see a lot of these patients but the ones that get sent to me, they get sent because they have PAH. So, acknowledging that they tactically fall under Group 5, when I see the patients, I say, you're a lot more like a Group 1, you've got a mean PA of 50 and a low cardiac index and a PVR of 10, you're a lot more like a Group 1. Or you're a lot more like a Group 3, maybe you've had sarcoid and now you've developed a little bit of pulmonary hypertension and your mean PA is 30 and your PVR is 4.5, and you really look more like a Group 3 to me. And so that's how I bucket them, in practice. And then I treat them accordingly.

You know, we have to be very clear like none of the therapies have been officially studied or indicated in these folks, but in clinical practice, I treat those former patients very aggressively like I treat the Group 1s and we've even had patients on parenteral prostanoids in that population. And the Group 3s are the ones who fall more like that. I tend to use, perhaps, more of an inhaled prostanoid.

Do you guys approach it the same way or anything to add to my simple view of the sarcoid patients?

Dr. Rajagopal:

No, I think you're right. If one of these patients ends up in your hospital in the ICU with right heart failure, due to PAH, trying them on parenteral prostanoids is very reasonable. And sometimes you can see amazing responses. Usually, you know pretty quickly whether it's working or not.

Dr. Preston:
Yes, the other thing is there are a couple of ongoing clinical trials focusing on sarcoid PAH that I recommend our colleagues refer their patients to.

Dr. McLaughlin:
Oh, that's a great point.

Dr. Preston:
So, we can learn more.

Dr. Rajagopal:
Yeah.

Dr. McLaughlin:
That's a great point.

Well, Sudar, Ioana, Rich, this was a really interesting conversation. It is a group of patients, you know, very complex, lots of different contributing factors. and that's probably one of the reasons that they're not included in clinical trials very often but important for us to address. So, thank you today for joining me and thank you. I hope you enjoyed our conversation on sarcoidosis patients and pulmonary hypertension.