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Case 3 and Panel- Right Heart Diagnostics in PAH

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Dr. Patel:
Victor really stole my thunder, because my case is also a case of Takayasu involving only the pulmonary artery. So, this case will be a little bit more kind of bread and butter, but just more of a case that emphasizes the importance of, you know, keeping an open mind and not anchoring on a diagnosis. But this is a 62-year-old female who I met when she was hospitalized for volume overload and congestion. She came in with a history of sickle cell anemia while she was followed at a practice that was outside of our purview, and she had a diagnosis of pulmonary hypertension. She was hospitalized with dyspnea, lower extremity swelling, and she had had a marked reduction in her functional capacity over the previous 2 years, and she was getting lightheaded with walking as well.

Her past medical history was notable for sickle cell anemia, as I mentioned. She obtained monthly exchange transfusions and, again, she had this diagnosis of pulmonary hypertension. She didn't know too, too much about the ways in which she was diagnosed with it, but she knew that she was on riociguat and she came in on that medication. She also had a history of atrial fibrillation and hypertension. You know, in talking with her, it doesn't seem like she had had a right heart catheterization prior to her initiation of riociguat.

On physical exam, she had an elevated JVP, she had a prominent C-V wave, her rhythm was irregularly irregular. She had abdominal distention, right-sided heart failure symptoms with 2-3+pitting edema of the lower extremities as well.

Here is her EKG. Her EKG demonstrated atrial fibrillation with a moderate ventricular response, notable absences on her EKG were absences of right ventricular hypertrophy, and again, we couldn't make much comment on her left atrial size or enlargement based off of her rhythm of atrial fibrillation.

Here's her transthoracic echocardiogram. The image on the left shows that she has severe left atrial enlargement. She's in AFib in this study as well. She had moderate TR on the right side, which is her RV inflow view. Shown here is her parasternal short axis view, there's some subtle flattening of the septum there. I mean, you might have to kind of squint your eyes for it, and that's at the mid papillary level. And shown on the apical 4-chamber view on the right, she has severe biatrial enlargement. That floating structure in the right atrium is her venous catheter for her monthly exchange transfusions. And her RV is mildly dilated, although compared to her RA, seems relatively small, and it's mostly annular in terms of the dilation, and her left atrium is severely enlarged, almost the size of her LV, if not similar in size.

So, here is her right heart catheterization, so her RA pressure. And this was after some days of diuresis, and then we were limited by a progressive AKI. Again, she's in Afib, A-flutter in this study, so somewhat difficult to determine, kind of, base of the A-wave. But her RA pressure was extremely high. Looks like it was, kind of, 25 or so. Here's her RV pressure kind of topping out at close to 60/20 with an EDP similar, kind of in the mid-20s or so. Her pulmonary artery pressure as well a PASP of about 63, 64. A mean PA pressure in the

mid-40s. And here's her pulmonary capillary wedge pressure, which demonstrated a wedge pressure of 34 or 35 or so. Her thermodilution cardiac output was 6.5, her thermodilution cardiac index was 3.1, her transpulmonary gradient was 10, and her PVR was only 1.5.

So, this was during her hospital stay on some IV diuretics. She had been maintained on riociguat, because we just didn't know much about her physiology at that point in time. And so, after we got her right heart cath and her numbers, we stopped the riociguat. And it's kind of an interesting phenomenon. What happened was we continued IV diuresis, but after topping the riociguat, she actually developed quite a bit of progressive and kind of profound AKI with IV diuresis, more so than what was occurring before. And, you know, potentially there was some thought that she was having some benefit in terms of maybe potentially some improvement of renal venous blood flow with the riociguat, and potentially stopping it quickly kind of could have initiated some of that AKI. Needless to say, she required a little bit of support from an inotropic perspective, mostly for the right side of the heart with milrinone, which was quite successful in approving her ability to diurese and she had a robust diuretic response. She was ultimately discharged on a look diuretic and an SGLT2 inhibitor.

She maintained clinical stability after –

Dr. McLaughlin:
Can I ask a question?

Dr. Patel:
Sure.

Dr. McLaughlin:
Where did her hemoglobin live?

Dr. Patel:
It was on the lower side, but with the exchange transfusions she kind of was 9 to 10. Yeah.

She was re-hospitalized 3 years later for heart failure. She had been able to maintain relative stability on loop diuretic and SGLT2 inhibitor. Had some issues with atrial fibrillation, but outside of that, no clinical decompensations until about 3 years later, in which she was hospitalized. A right heart cath was repeated demonstrating an RA pressure of 18, a PA pressure of 63/30 with a mean of 44, and a wedge of 26. Her index was 2.3 by thermodilution and she had a PVR of 3.9. So, as compared to her prior right heart cath, her PVR looked a little bit higher this time around 3 years later. She had developed a little bit more of a transpulmonary gradient and, you know, may have progressed from that isolated, you know, pulmonary venous hypertension to more of a pre- and post-capillary hypertension phenomenon.

She was initiated on sacubitril-valsartan and spironolactone, and that's kind of how she is now, quasi-stable, and we are evaluating her for clinical trials of combined pre- and post-capillary PH. PA denervation and levosimendan are on the possible dockets for her. But I brought this case up because this patient obviously had some risk factors for pulmonary hypertension for sure, risk factors for PAH given her sickle cell anemia and multiple ways in which sickle cell can cause PAH, and potentially there was some anchoring bias based off of her history of sickle cell, but also had developed, independently of that, multiple other risk factors, atrial fibrillation, hypertension, and I didn't mention she was obese as well.

Her atria developed quite a bit of remodeling in the setting of her atrial fibrillation. It seems that that alone was contributing a lot to her left-sided heart failure as well. But kind of an interesting case again, one where the hemodynamics really tell the true story, and I thought an interesting case just to bring us back, similar to Dr. Raza's, of the importance of looking at invasive hemodynamics to kind of get a true sense of physiology. A lot of times with these patients as they're getting older, they don't live in a vacuum and they develop multiple risk factors, and so it's important to always take a look.

Dr. McLaughlin:
You know, it's really interesting that her cardiac output wasn't higher, right? Like, I don't see that many sickle cell patients, but many of the sickle cell patients I've seen, they develop basically high-output failure, like their wedge is high because their cardiac output lives at 10, 12, you know, whatever, because they're anemic. So, I find it really interesting, like how she developed such severe, you know, both left heart and pulmonary vascular disease with a cardiac output living in the normal range as a sickle cell patient.

Dr. Patel:
Yeah. Yeah, I agree, and you know, we're still learning a little bit about her history of sickle cell or if it had been managed well from an exchange transfusion perspective that she never had such severe anemia, that she never developed that high output state, and then she just kind of developed these co-morbidities that kind of tipped her into left heart failure and PH. But, yeah, we were surprised by that

as well and we have a fair amount of our patients with sickle cell that come through our HFpEF clinic with high-output failure as well.

Dr. Cuttica:
I think the other interesting thing to think about in a sickle cell patient is does she have a history of recurrent acute chest syndrome, and how many sickle crises does she have, right? Because the chest syndromes are associated with increased thrombotic risk and chronic clot have been described in patients with sickle cell anemia, so it would be something to look for to make sure that she doesn't have some degree of obstruction that might benefit from a BPA or something like that. Rarer in a sickle cell patient, but definitely something that happens. And then, the fact that she's 62 years old and has sickle cell anemia itself is pretty interesting, right? And in that, sort of somewhat controversial initial description of PH in sickle cell anemia, right, ultimately it is the older patients that survive into older age that are the ones that truly develop the vasculopathy where they act like group 1 PAH, where you would want to maybe think about PAH-directed therapies. But such a complicated patient population because of all the different ways they can develop PH. It's interesting.

Dr. Patel:
Yeah, and I agree. We did look into, with the nuclear scan, to just kind of cover our bases to see if she had any V/Q, you know, mismatching.

Dr. Cuttica:
Yeah, especially with that line that's line that's chronic indwelling.

Dr. Patel:
Right, exactly. Dr. Raza? Go ahead.

Dr. Raza:
Okay. I was going to ask if she ever had a cardiac MRI? You know, her echo was really striking. Did she have like, you know, it looked like maybe she had some LVH, and like, with all of her transfusions, like did she ever have a high ferritin and evidence of iron deposition disease within the myocardium? And then you also mentioned that she was obese, and I know that we've certainly seen some patients who especially have a lot of pericardial or epicardial fat, that they get, sort of like that increased pericardial restraint and get that restrictive hemodynamic picture, which it looked like she had, especially on that first right heart cath, those tracings that you showed with like the nice prominent Y descent and so on.

Dr. Patel:
No, those are great points. So, she did ultimately get the MRI and she did not have any evidence of hemochromatosis or, you know, given those kind of owl-eye biatrial enlargement, we just wanted to, you know, make sure as well that she didn't have cardiac amyloidosis as well, which she did not. But it's a great point with regard to her obesity. She does have an epicardial fat pad, there's probably some interventricular dependence that was definitely present on that first echo as well. She would benefit from weight loss, and potentially that physiology may improve with therapies to improve her weight. All great points though.

Dr. Moles:
Ravi, I'm going to ask a question that always perplexes me when I'm in the wards taking care of patients on the cardiology floor. Why do you think some patients like this, that are clearly hyperkalemic by exam, by hemodynamics, you try to diurese them, and you can't? And I found very interesting that you chose milrinone when the cardiac output was still kind of within a very reasonable, kind of normal range.

Dr. Patel:
Yeah. Well, I think in her, and in many of these patients, the kidney is underappreciated in terms of its overall health with the ability to diurese. And with her, I think that there was some renal malperfusion that occurred after stopping the riociguat. You know, there's this – of therapies to treat heart failure, there are very few that have shown benefit to kidney function. A lot of times we have to accept in some of our heart failure therapies that the kidneys may take a hit. One of the therapies that does benefit the kidneys is sacubitril-valsartan, and sacubitril is a compound that can dilate the afferent arteriole of the glomerular bed and that dilation, I think, improves overall renal blood flow. Potentially something like that was happening with the SCG stimulator, and that when we took it away, the renal arteries kind of clamped down causing this AKI with diuresis and she needed some other inodilator to kind of improve that, but I think there's some crosstalk there with the renal syndrome that is underappreciated with our patients that have a lot of diuretic resistance.

Dr. McLaughlin:
Great. Alright, thank you. Well, we've reached the end of our program, and I think I want to just start out by thanking the faculty. We had a number of, you know, really wonderful talks, a lot of great discussion, and really appreciate the expertise that everyone brought to this and thank you all for taking such a big chunk of your Saturday to be with us. So, I really enjoyed today and appreciate you all. And I'll let

Mike say some final comments as well.

Dr. Cuttica:
I agree. Thank you all for coming. Thank you, Val, for cohosting this with me, and yeah, thank you for coming and spending the day with us. I also really enjoyed this. The faculty all did a great job. I really, as usual, learned a lot from all of you. So, thank you.

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