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Case 2 and Panel- CTEPH and PH

Announcer:
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Dr. Mole:
I'm going to share with you a case of a CTEPH patient and I think it will generate a lot of good discussions.

This is a 21-year-old female who doesn't have a history of significant comorbidities who presented to the Emergency Department with worsening dyspnea and hemoptysis. And when you go into her past medical history, the only thing that we could get out of her was that 2 years before presented to the emergency department, she was told she had a pneumonia, after which she was treated with antibiotics but had persistent dyspnea since then, which has been progressing over time. Her previous workup was unremarkable, and the exertional dyspnea was attributed eventually to anxiety. There is no personal or family history of venous thromboembolic events, and when in the emergency department, she has a CT angiogram concerning for pulmonary embolisms and an echocardiogram consistent with severe pulmonary hypertension.

So, I'll show you some of the tomography results. You'll see first that the main pulmonary artery is dilated, and you'll see some severe, kind of, narrowing of the left pulmonary artery very proximally. You see some degree of air space disease as well. When you look at the vasculature of the lung, you see that the right seems a little bit more hypovascular and there really is no vessels that we can see, there's no contrast being seen on the left lung. When we look at the right pulmonary artery, kind of in the mid segment, right away there's a severe stenosis again. And when we look at the coronal cuts, you'll see that there's a lot of collateral around the pulmonary artery. So, that's a little bit more of the findings.

I'll show you the lung windows. We can see here on the left that the left lung is very hypo vascular compared to the right one. And on the right picture you'll see that the left lower lobe has an area of parenchymal density that we attributed to probably a pulmonary infarct.

These are the echocardiographic findings. In the partial lung axis view, the patient is tachycardic. You'll see that the right ventricle is severely dilated. In the short axis view you see the interventricular septum being flat in diastole and in systole, as well the left ventricle is very small compared to the right. You'll see that presence of RVOT Doppler notching and short acceleration time. You'll also see the right ventricle in the 4-chamber views being very dilated and dysfunctional. The TR peak velocity is estimated as being severely elevated and the inferior vena cava is dilated and nondistended.

So, consistent with severe pulmonary hypertension. I think that we can agree that there are many findings that makes us think that this is pre-capillary. After discussion we have a high suspicion that this is not an acute pulmonary embolism, but rather that this has been present for some time. We ordered a V/Q scan and just to show you that the left lung has decreased ventilation, especially in the left lower lobe. In that area there was sub parenchymal abnormalities, but there is a very significant in perfusion on the left lung compared to the right, and I want you to kind of remember this picture because the right lung appears quite normal, but it my not be the case.

We proceed to further evaluation of CTEPH. We do this pulmonary angiogram. This is the right lung in a frontal view. You see in the mid segment this severe stenosis that we saw on the CT angiogram, and also, we see decreased perfusion throughout the entire left lung. In the oblique view, we see that severe narrowing, but also more severe distal narrowing as well.

This is the left lung, just to give you – the main pulmonary artery is just this segment right here. It is severely obstructed. There's actually no perfusion whatsoever to the left lower lobe and this is the oblique view that shows that there is severe stenosis and also total occlusions.

These are the hemodynamics. The right atrial pressure is 16 and that's severely elevated. The right ventricular pressure is 110/11 with an EDP of 20. The pulmonary artery pressure is 111/47 with a mean of 71. The wedge was normal at 12. The cardiac output was 2 and the cardiac index was 1.3, and the pulmonary vascular resistance was 30, and I have to say that's not the highest that I've seen.

So, we discussed the case in our multidisciplinary meeting. We agree that something doesn't completely fit the picture and we tried to look for those zebra diagnosis. Our radiologists go through the CT pulmonary angiogram, and they say there's nothing else going on, this is CTEPH, and we decide to take the patient for a pulmonary endarterectomy. John reviewed the images, and they said this is pretty proximal disease, I'm going to be able to make this patient feel better because I'm going to reach out and remove a lot of clot. But once John gets into the pulmonary arteries, there is no clot. And what he finds is that there's multiple stenosis and occlusions that are noted that appears to be a constriction phenomenon rather than intravascular obstruction, and there's a fibrotic appearance to the arteries. What you can see there is my reaction when I heard what happened and this is probably how John felt, more like Captain Sully taking control of the aircraft without engines. And what John did was take samples and the patient was transferred to the CVC ICU and actually did pretty based on our concerns that because John was not able to remove clot, the pulmonary vascular resistance basically remained unchanged and the possibility of an acute RV failure that may be, you know, very severe, that didn't happen.

The patient went to the floor on day 4. We consulted basically every single specialty in the hospital, and we had a CT of the neck, chest, abdomen, and pelvis with contrast, and we looked at every single other major artery and we did not find any other evidence of stenosis. So, the biopsy results come back, and it shows a granulomatous vasculitis consistent with isolated pulmonary artery Takayasu arteritis. The patient is given high dose steroids and infliximab in the hospital and then is transitioned to oral steroids and methotrexate as an outpatient. And this is the diagnosis, isolated Takayasu arteritis of the pulmonary arteries.

So, those of you who are not familiar, Takayasu arteritis is a large vessel vasculitis which disproportionately affects young women in their second or third decades of life and more predominantly in the Asian population. There is no gold standard test to diagnose Takayasu arteritis. The clinical assessment and labs correlate poorly with disease activity. The pulmonary artery involvement is actually somewhat common in Takayasu arteritis and occurs somewhere between 20 to 56% of the patients, but isolated Takayasu of the pulmonary arteries is extremely, extremely rare. Immunosuppression in general is the mainstream of treatment for Takayasu arteritis. And pulmonary angioplasty has been described in small series to improve hemodynamics and functional capacity, but the consequences are a very high rate of complications that's been described above 30%.

The clinical course, we repeated a right heart catheterization and pulmonary angiogram after 4 months of immunosuppression. The truth is that the hemodynamics didn't significantly change. The pulmonary anatomy did not significantly change. We proceeded with optimization of the hemodynamics. We started tadalafil and macitentan. The patient significantly improved in symptoms and RV function. The right ventricle is still mildly dysfunctional and we're planning to start parenteral prostacyclins. We've been discussing with multiple colleagues around the globe and the truth is that there really is no consensus on how to treat this. And we are considering in the very early phases to do balloon pulmonary angioplasty once the hemodynamics are optimized.

Just want to bring up a few differential diagnoses when you're thinking about CTEPH cases. CTEPH, when we talk about it, we're presuming that the obstruction is related to blood clot organized thrombi, but there's a lot of other medical conditions out there that can cause vascular obstruction of the pulmonary arteries. I'll have you think about pulmonary artery sarcoma, especially when the disease is very unilateral, and I will say that one of the tests that is more helpful in this condition is getting an MRI of the pulmonary arteries that would show that that lesion is vascularized and has significant enhancement compared to a chronic clot that should not be significantly vascularized. We had a patient also who we diagnosed with a pulmonary artery angiosarcoma after having a negative PET scan and there are intimal sarcomas that tend not to light up on a PET scan. So, a cardiac MRI is a very helpful test.

The other thing that I'll make you think about is atrial septal defects, or any conditions that can increase right-sided flow that can dilate the pulmonary arteries and can create that in situ thrombus. In these patients, of course, you don't want to take them to have a pulmonary endarterectomy right away because the problem is not a pulmonary vascular disease. It's not a mechanical obstruction, it's rather that extra flow through the ASD. And with that, I'll go to the conclusions.

CTEPH is a complex medical condition characterized by chronic emboli causing mechanical obstruction to the pulmonary blood flow.

Thrombus characterization is really difficult clinically. Not every pulmonary vascular obstruction is thrombus, so keep an open mind. Takayasu arteritis can affect the pulmonary arteries. Systemic arteries are usually involved. Isolated Takayasu arteritis of the pulmonary arteries is very rare and truly this is a multidisciplinary approach where we need all our colleagues to be involved in chiming in and helping us diagnose these patients and treat these patients. Thank you.

Dr. Cuttica:
Great case. You made me feel a lot better about myself because one of the worst clinical experiences I've had in the last 5 years was being called into the operating room where our surgeon said, hey, there's no clot in this lady's lung. It ended up being vasculitis. But I think, you know, that was really a great case, and I loved the cartoon, that's exactly how you feel when that kind of thing happens. But the imaging was so striking when you put that up there. The degree of stricture, and it was into the lower lobes, right? It was, you know, make you think CTEPH, but without seeing the intramural thrombus or the webbing, it was such prominent strictures. You know, it would be a unique CTEPH case had it been CTEPH, but that was really striking. I thought that was a really interesting case. Thank you.

Dr. McLaughlin:
Yeah, it is. This is my patient, right? Yeah. I remember the day that Jonathan called me when he operated. I mean, both of our stomachs sunk. And you know, in reality, if you don't do this once in a while, you're not doing enough endarterectomies. You know, it's just like an appy, and the goal is, you hope everyone learns. And I think about what I might have done differently in her case, and, you know, maybe I'll be more attuned to it if this happens again, very rare if this happens. And you know, I don't know if MRI would be useful or if we would get inflammatory markers, or that sort of stuff. You know, it was very interesting because it's unusual to have it isolated to the pulmonary vasculature. Like, no other artery was involved. But on the other hand, that's really good for her because while she's gotten a little better on oral therapy, like, generally these patients don't respond long-term to pulmonary vasodilators and probably, if she were to get worse, like she would still be a lung transplant candidate because it doesn't involve any other vessels. So, that's like the one, you know, glass half full type of thing for her.

Dr. Cuttica:
Yeah, I agree. You know, those mimics that you look at that we struggle with in that multidisciplinary meeting that we all have looking at those can be really hard. And you're right, ultimately in our case looking back in retrospect, right, we came back and said our patient's lesions were predominantly bilateral upper lobe. Like, she had no perfusion to the bilateral upper lobes, maybe a little bit of changes in the bases, and we went back and forth about, well, that's odd for CTEPH to have only upper lobe disease and not lower lobe disease. MRI'd her, and ultimately we said we've got to give her a change, we think this is clot, we can't rule it out. The other big mimic that we struggle with is the fibrosing mediastinitis patients where they come in and you're like, those perihilar findings, are they intravascular clot, are they compression? It can be really challenging sometimes. And not only within the institution, sort of all the groups coming together, but like you said, reaching out to colleagues and saying, what do you really think of this, before you take the patient to the operating room is important.

Announcer:

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