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Diverse Manifestations of Sickle Cell Disease

Announcer:

Welcome to CME on ReachMD. This episode is part of our MinuteCME curriculum.

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Dr. Andemariam:

This is CME on ReachMD, and I'm Dr. Biree Andemariam. Here with me today is Dr. Caroline Freiermuth. We will be discussing the diverse manifestations of sickle cell disease through brief case vignettes. Dr. Freiermuth, please present case vignette number 1.

Dr. Freiermuth:

So we have a patient named Marsha. She's a 31-year-old female with a history of hemoglobin SC [sickle cell] disease. She presents to the emergency department complaining of pain in her chest and her right hip. This started last night. She slept rather fitfully because of the pain. She did take her regularly scheduled 20 mg of OxyContin yesterday evening and again this morning at 9. And then further she took rescue doses of 10 mg oxycodone at 3 AM and 7 AM. However, she's had minimal relief, and she's complaining of pain at a 9 out of 10. She says that she has no trouble breathing. And she just complains of a lack of energy.

And so when I approach this patient in the emergency department, I really want to keep in mind that a vaso-occlusive episode, although it is the most common reason for presenting to the emergency department, is a diagnosis of exclusion. I must always think about ruling out other bad comorbidities. And realize that many complications associated with sickle cell disease happen during acute painful episodes. One of the things that catches my attention is her complaint of chest pain, and I really am thinking acute chest syndrome and what more do I need to rule that in or out. This accounts for a significant number of deaths in sickle cell disease. And so anytime somebody complains of symptoms such as chest pain, cough, difficulty breathing, fever, or I noticed they're hypoxic, I really need to screen further and think about whether or not I need that chest x-ray.

And Dr. Andemariam, I'll let you present case 2.

Dr. Andemariam:

Thank you so much. Alicia is a 30-year-old woman with hemoglobin SS disease. She has had multiple complications, including the acute chest syndrome requiring ICU stay and brief intubation, splenic sequestration crisis which required a splenectomy, and sickle retinopathy with retinal detachment. She has had frequent pain crises as a child but was able to persevere through school and is now a practicing attorney. She is recently married and is contemplating having children. Her ancestry is Nigerian and her husband is Italian. They present to discuss the risks of pregnancy as well as the risks of having a child with sickle cell disease.

So Dr. Freiermuth, please start the conversation about these two cases.

Dr. Freiermuth:

Well, you know, I think it's wonderful what you get to see in the outpatient world, people advocating for themselves and coming in asking you about, you know, what the risk might be for their chances of having, you know, a child with sickle cell disease. And we in the emergency department really, you know, try to push patients to advocate for themselves as well, educating them that when they

present, they really need to make it clear that they have sickle cell disease. So you can't just come in and say, 'I'm having hip pain,' but you really need to say, 'I'm having hip pain, consistent with my sickle cell disease,' or, 'I'm having a vaso-occlusive episode.' And that really helps our triage nurses know, okay, somebody has, you know, a disease process that puts them at much higher risk for morbidity and mortality than the general population who might come in complaining of hip pain, and helps us triage them appropriately and get them back quickly.

Dr. Andemariam:

I 100% agree with you. And I think your case highlights the importance of always keeping in mind that when a patient comes in for an acute pain crisis, they also may be harboring early signs of the acute chest syndrome, which we know can be fatal in many circumstances, particularly when it's not recognized early.

And I think another thing, you know, I agree with you regarding the case that I presented about Alicia. You know, Alicia is living her life. She's got her graduate degree, she's married, she's contemplating her future. And I think a lot of times when we see patients in the acute setting, we forget that they have a life and they have dreams and they have ambitions. And I think that's important to keep in mind even though we don't always in the acute setting, get to see them in that ongoing fashion, like I do in the outpatient ambulatory setting.

Well, this has been an eye-opening discussion. I would like to thank Dr. Caroline Freiermuth for joining me today. Unfortunately, our time is up. Thank you for listening.

Announcer:

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