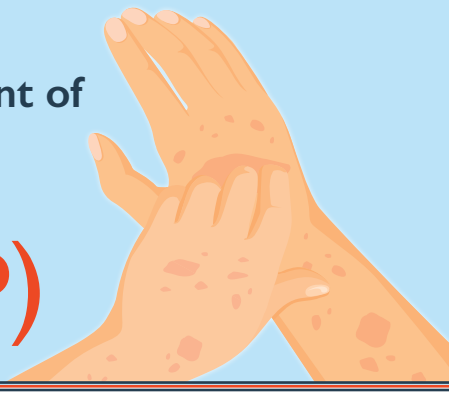
Improving the Understanding, Diagnosis, and Management of

Generalized Pustular Psoriasis (GPP)



Podcast Series: Episode 2

Adrian: Hello and welcome to "Improving the Understanding, Diagnosis, and Management of Generalized Pustular Psoriasis," a podcast series brought to you by the American Academy of Physician Associates and the France Foundation. This activity is supported by an independent educational grant from Boehringer Ingelheim. This is episode two of a five-part series focused on generalized pustular psoriasis or GPP. In this episode, we will discuss the diagnostic challenges associated with GPP.

And we'll provide a deep dive into screening protocols, especially those surrounding distinguishing GPP from other skin conditions. We're going to talk about recognizing related comorbidities and assessing severity over time. My name is Adrian Banning. I'm a PA and an associate professor in the PA program at the Delaware Valley University PA Program in Doylestown, Pennsylvania.

I'm joined today by my wonderful colleagues, dermatology PA Terri Nagy and dermatology NP Leigh Ann Pansch, as we discuss generalized pustular psoriasis. Leigh Ann and Terri, thank you so much for being here. Will you each tell us a little bit more about yourselves?

Terri: I'm Terri Nagy and I am a physician assistant. I've been working in dermatology for 20 years now and as a practicing PA for 25, which is crazy. I'm in Colorado Springs right now, but I spent most of my time working in Ohio prior to moving to Colorado about three years ago.

Leigh Ann: And I am Leigh Ann Pansch. I'm a nurse practitioner practicing in the private practice setting in Cincinnati, Ohio. I've been in practice for just shy of 15 years. And I absolutely love treating patients with inflammatory skin disease.

Adrian: Terri, Leigh Ann, I love being here with both of you because you're both so passionate about dermatology and talking about GPP. It's not a room I find myself in every day, but to talk about this topic, I can't think of two better friends to be here with. As we jump in, Leigh Ann, can you talk to us about some of the common ways a patient could present to their primary care or dermatology office when they have GPP?

Leigh Ann: Absolutely. I think first and foremost, this word generalized in front of generalized pustular psoriasis, it means really that this is a full body systemic condition. And I think just recognizing that these patients are really sick, that they often have some key findings, including a history of maybe some generally mild plaque type psoriasis. They may have had a recent illness or have recently been on systemic glucocorticoids. They often have significant pruritus, and they will describe this as the worst ever just prior to the onset of the rash.

And it's important to examine every skin surface. I always say every nook and cranny to look for these monomorphic pustules. And monomorphic here, it means uniformity in size and in shape. Patients with generalized pustular psoriasis can be erythrodermic and they may present with significant and acute inflammatory arthritis, which is so debilitating that it affects their activities of daily living.

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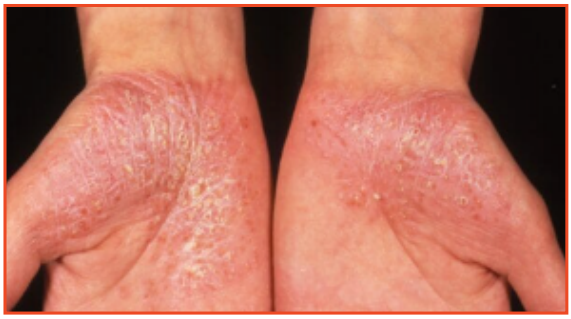

Figure 1. https://www.rarediseaseadvisor.com/features/recognizing-managing-flares-generalized-pustular-psoriasis/stular-psoriasis

Leigh Ann: They may have a low grade fever, report some anorexia, maybe have some difficulty sleeping due to all of the pain and inflammation. They may also be hypocalcemic. To summarize, generalized pustular psoriasis presents acutely and abruptly.

Adrian: That's so good to know. So this is not someone just coming in to say, "Hey, I just found this new thing on my skin. What do you think?" This is someone who's really sick. It's come on all of a sudden and it's a big presentation. There are some other things that might sound like that, a provider, a clinician, a PA, an NP, whether you're in primary care or derm, they might have a few differentials in mind if they hear about those symptoms or see those signs on exam.

So, because there might be a couple differentials, Leigh Ann, what can we do to make sure that we're getting the right one?

Leigh Ann: That's such a great question. I think the truth is we may consider generalized pustular psoriasis part of our differential diagnosis, especially if we see a pustule. But the way that we confirm diagnosis is typically with a punch biopsy completed for histopathology.

Adrian: So just take a biopsy of one of the lesions that you see and is one enough?

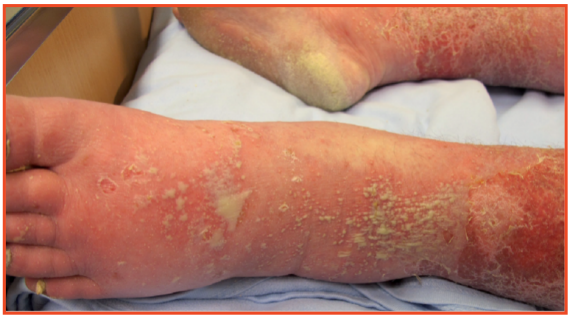

Figure 2. https://www.medicaljournals.se/acta/content/html/10.2340/00015555-2467

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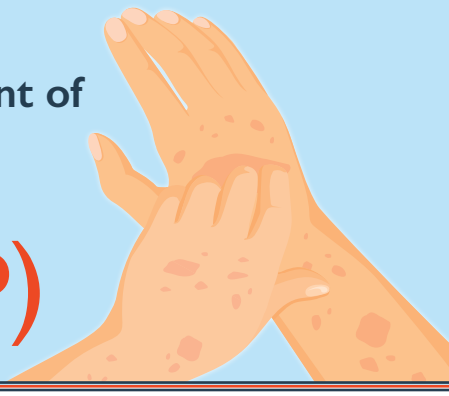

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Leigh Ann: I absolutely think one is enough. I will tell you that I likely would recommend a punch biopsy in this case, at least a three millimeter. And I would not do a deep shave because I actually want to see what's happening between the layers of skin so that I can see those pustules forming.

Adrian: Good to know. Thank you so much, Leigh Ann. Terri, so now that we've talked about how to get to the right diagnosis when we have GPP as an option. What about the other illnesses that a person might have? Are there any regularly co-occurring comorbidities? How would we recognize them if there are?

Terri: Adrian, that is such a great question. I think when we talk about the idea of comorbidities, we have to think about things that trigger pustular psoriasis. We know that in some patients they have preexisting plaque psoriasis, but that's not always the case. So we have to think about what medications a patient's on, because those could be the triggers and what diagnoses are associated with those medications.

We have some patients on TNF-alpha inhibitors who are at risk for generalized pustular psoriasis. TNF-alpha inhibitors are certainly used for plaque psoriasis, but they are also used for other autoimmune disorders. Diagnoses such as psoriatic arthritis and even inflammatory bowel disease to name a few. So those can be comorbidities for our patients with generalized pustular psoriasis. Cyclosporine is also a trigger.

It is used in transplant patients and also in patients with rheumatoid arthritis and psoriasis. So those are also other comorbidities for this disease state. A study published in the Journal of American Academy of Dermatology in October of 2022 found that individuals with GPP actually had a higher incidence of hypertension, Type 2 diabetes, and hyperlipidemia.

It also noted that women were more likely than men to develop GPP, and the average age in years was in the mid-fifties. So, there are definitely indicators for disease risk.

Adrian: That's a lot, Terri, because some of the things that you listed have a lot of burden associated with them already. So to think that you could have rheumatoid arthritis and GPP or hidradenitis suppurativa and GPP, that's a lot for a person to go through and to think about that compounded burden. Thank you so much.

Terri: Of course.

Adrian: Leigh Ann, it sounds like maybe patients with GPP might be a group who get used to their symptoms changing over time, maybe getting worse, maybe having some comorbid symptoms too. Hopefully we help them improve, but any process, of course is dynamic with ups and downs. Will you talk to us a little bit about how we can assess GPP severity over time?

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Leigh Ann: Absolutely. To evaluate the extent of disease, we often use body surface area or BSA analysis. In this sense, a patient's own hand is about 1% of their body. Remember, it's their hand, not yours. You can also use the rule of nines to evaluate the extent of disease. And remember, the rule of nines comes from our assessment and evaluation of burn victims. It's really important to classify the body locations affected, the look and feel of the plaques. Are they red, scaly, or thickened?

We definitely want to evaluate for joint pain. We also want to get our hands on and actually palpate and examine those antitheses and joints. And then we want to measure the disease quality of life. In the world of psoriasis, it's also important to identify when special sites are involved. This is the face, the scalp, the hands, and the genitals. When these areas are involved, we recognize that there is greater impact on quality of life. And then lastly, I always try to ask a really simple question.

If your generalized pustular psoriasis were improved, what would you be doing more of that you aren't able to do now? And then I want to include a patient's direct quotes in the chart as much as possible.

Adrian: These were amazing tips. A reminder about the rule of nines, using the person's own hand, and just really asking them so directly, what is being limited in your life? What would you be doing more of? What an easy and simple way to really assess the severity of someone's disease. Thank you so much, Leigh Ann.

We've learned so much about diagnosing GPP in this episode. Terri, would you be so kind as to summarize some of that for us? What are your key takeaways that we talked about today?

Terri: Of course, I'd love to do that. I think one of the most important points is to remember generalized pustular psoriasis can be induced by medications, and oral steroids can be the biggest culprit. GPP patients can be acutely ill, and many will present with erythematous, pruritic patches, and plaques, which soon evolve into those monomorphic pustules that Leigh Ann described for us earlier.

It's really important to perform that complete head to toe skin exam, including an evaluation for joint pain. GPP can have a huge impact on a patient's quality of life. So making sure we're cognizant of that is important when moving forward with any treatment plan.

Adrian: Thank you both so much, Terri and Leigh Ann. That was a fantastic overview of the challenges that dermatology and primary care providers might face when screening for and diagnosing generalized pustular psoriasis. What a serious condition so important for all of us to know more about. Thank you for listening to this episode of "Improving the Understanding, Diagnosis, and Management of Generalized Pustular Psoriasis."

Please tune in to the other podcast episodes in this series where we talk through the pathophysiology of GPP, the steps to treatment, current and emerging treatments, and, finally, implications for patient's quality of life related to GPP. You can find the full list of podcast episodes in AAPA's Learning Central catalog at cme.aapa.org.

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