Don't Be A Chicken: Uncovering A Rare Case of Breakthrough Chicken Pox

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Background

- Chicken pox, or primary varicella, is a contagious illness caused by varicella zoster virus (VZV), resulting in a pruritic rash among other symptoms.
- Before vaccination was available, 4 million people contracted chicken pox each year in the US, resulting in 10,000+ hospitalizations and hundreds of deaths.
- Vaccination has significantly reduced chicken pox infections, although vaccinated patients are still at risk to develop herpes zoster infections and, rarely, breakthrough primary varicella infections.

Patient Presentation

- A 39-year-old man with no medical history presented with one week of progressive diffuse pruritic rash with no pain, subjective fevers, and fatigue. He received all childhood vaccinations including chicken pox (confirmed by family).
- **Physical exam** was notable for maculopapular rash with central hemorrhagic crust on his face, trunk, back, and extremities, with one pustule on his right thigh.
- Labs including CBC, CMP, TSH, and ESR/CRP were within normal limits and blood cultures showed no growth at 48 hours. HIV and RPR were negative. **VZV PCR of lesions was positive.** HSV PCR and Monkeypox PCR of lesions were negative.

Rash on back (1)

Right thigh pustular lesion (1)

Rash on lower extremities (1)

Clinical Course

Hospital Day	Event
Day -7	Patient visited urgent care clinic one week prior to admission with rash on his back. Patient was given oral prednisone course and oral hydroxyzine as needed for itching.
Admission	Patient presented to emergency room with progressive rash. Workup was initiated and IV acyclovir was started. Dermatology was consulted. Patient was placed on airborne and contact precautions.
Day 2	VZV PCR returned positive while HSV PCR and Monkeypox PCR was negative. One new vesicular lesion on left thigh was noted.
Day 3	No new lesions were noted, and patient was discharged with PO valacyclovir three times per day for 7 days. Patient was determined to have a breakthrough primary varicella infection.

Chickenpox Overview

- Chicken pox is spread through direct contact with a rash or bodily fluid, surfaces, and respiratory secretions.
- Patients typically develop a pruritic maculopapular rash with vesicles/pustules that scab over to form crusted lesions. The rash may start on the chest, back, or face, then spread to extremities and sometimes the mouth and genitals.
- Some will have systemic symptoms including fever, myalgias, fatigue, and/or respiratory symptoms. Illness typically lasts 7-10 days.

Primary Varicella (2)

Herpes Zoster, (3)

Herpes Zoster (3)

Disseminated Herpes Zoster (4)

Chickenpox Vaccinations

- CDC recommends two doses of chickenpox vaccine for children and adults who have never had chickenpox and were never vaccinated. The recommended schedule is one at age 12-15 months and one at age 4-6 years.
- The two vaccines offered, Varivax and Proquad, are live attenuated varicella-zoster vaccines, which induce both humoral and cell-mediated immune responses. The duration of protection is unknown.

	One dose	Two doses
Effectiveness	• **82% effective at preventing any form of varicella** • **Almost 100% effective at preventing severe varicella**	• **92% effective ay preventing any form of varicella** • **Almost 100% effective at preventing severe varicella**

- A systemic review of breakthrough infections involving 34 published articles found only **60 cases** of severe breakthrough infections with disseminated rash and involving organs other than skin. The majority of all breakthrough infections occurred in people who had received 1 dose vaccinations.

Primary Varicella vs. Herpes Zoster

It requires clinical reasoning to differentiate between primary varicella and herpes zoster as both lesions will test positive for VZV PCR.

Primary Varicella (Chicken Pox)	Herpes Zoster (Shingles)
• **Etiology: VZV (DNA virus)** • **Breakthrough varicella** is an infection with VZV in a vaccinated person more than 42 days after vaccination, typically appearing with less than 50 lesions. • **Appearance:** vesicles which morph into scabs starting in chest/trunk and spreading all over body. **Typically not painful.** • **Complications:** skin infections, pneumonia, and encephalitis. • **Treatment:** No routine treatment for children under age 12. Immunocompetent patients at high risk for complications receive oral therapy (valacyclovir or acyclovir). Individuals with complications receive IV acyclovir.	• **Etiology: reactivation of VZV**. After a person acquires VZV, it can remain hidden in the dorsal root of spinal nerves which can reactivate years later into herpes zoster. • **Appearance**: vesicles which morph into scabs typically in dermatomal distribution. **Often with pain.** Disseminated zoster can occur in immunocompromised patients. • **Complications:** post-herpetic neuralgia, herpes zoster ophthalmicus, Ramsay Hunt syndrome, encephalitis • **Treatment:** Oral therapy (valacyclovir or acyclovir) for all patients within 72 hours of symptoms. Patients who are high risk receive oral therapy after 72 hours of symptoms. Patients with high risk for dissemination should receive IV acyclovir.

Discussion

- ***This is a rare case of severe breakthrough primary varicella in an adult.***
- Our differential included breakthrough primary varicella, disseminated zoster, HSV, monkeypox, syphilis, and drug rash. Lesions were positive for VZV PCR which narrowed the case to primary varicella and herpes zoster.
- Given the diffuse rash in an immunocompetent individual with pruritis and no pain, we determined our patient had breakthrough primary varicella. This is atypical given our patient had a disseminated rash rather than a rash with under 50 lesions.
- This case highlights the importance of keeping a broad differential and shows how vaccinations do not always fully prevent disease.

Learning Points

1. **Explore effectiveness of chicken pox vaccinations and the possibility of breakthrough infections.**
2. **Understand differences between primary varicella and herpes zoster infections.**

References

1 Photos of patient in this case.
2 Photo obtained from: https://www.nhs.uk/conditions/chickenpox/
3.Photo obtained from: https://www.verywellhealth.com/what-is-herpes-zoster-3132938 `
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7 Leung J, Broder KR, Marin M. Severe varicella in persons vaccinated with varicella vaccine (breakthrough varicella): a systematic literature review. Expert Rev Vaccines. 2017 Apr;16(4):391-400. doi: 10.1080/14760584.2017.1294069. Epub 2017 Feb 28. PMID: 28276305; PMCID: PMC5544348.