

CASE REPORT

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# Atypical, generalized cutaneous varicella zoster in a patient on chemotherapy for leiomyosarcoma

Ashley N. Elsensohn, Maryam Liaqat, Dante Garcia, Charay Jennings-Dover, Gregory Simpson

## ABSTRACT

**Introduction:** Most cases of atypical generalized varicella zoster virus (VZV) have been reported in patients with post-renal transplantation or hematogenous malignancy. **Case Report:** We present a unique case of atypical, generalized cutaneous VZV in a solid tumor patient. Of interest, our patient was immunocompromised secondary to chemotherapy for a stage IV leiomyosarcoma. **Conclusion:** This case highlights the diverse presentation and subsequent challenges in the diagnosis of generalized zoster, which carries a high mortality in elderly and various immunocompromised populations. The case briefly reviews diagnostic and therapeutic recommendations, as well as complications of generalized zoster. We recommend a high level of clinical suspicion and a low threshold for treatment of suspected disseminated zoster in immunosuppressed, solid tumor patients.

**Keywords:** Atypical VZV, Cutaneous VZV, Generalized Zoster, Varicella

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## INTRODUCTION

Atypical generalized varicella zoster virus (VZV) is a term used widely in literature to describe diffuse, varicella-like skin lesions, without primary dermatomal involvement [1]. Atypical generalized zoster is seen almost exclusively in immunocompromised patients. Cases in literature describe patients with post-renal transplantation or hematogenous malignancy[2,3]. Our patient presents a unique case of atypical generalized zoster in a solid tumor patient immunocompromised secondary to chemotherapy for stage IV leiomyosarcoma.

## CASE REPORT

A 53-year-old Hispanic female with a history of stage IV leiomyosarcoma presented with a one-week history of worsening neck and shoulder rash. She had undergone palliative chemotherapy with Adriamycin 10 days prior. She first noted the rash approximately three days after chemotherapy and described solid papules on the right side of the neck that had progressively grown in size, with additional vesicles appearing on her right shoulder.

She reported pain and tenderness over the lesions but denied pruritus. Initial lab values included WBC 500 (Differential: neutrophils 15, bands 2, lymphocytes 41, monocytes 36, eosinophils 2, basophils 4), Hb 8.2, MVC 91, platelets 22,000. Basic metabolic profile was within normal limits. The patient was admitted to the hospital for neutropenic fever and started on cefepime and vancomycin for suspected cellulitis.

On examination, there was a confluent, violaceous plaque with an erythematous base that extended from mid-chin to right posterior neck, scalp, and clavicle. The rash did not cross the midline and had yellow scaling at the borders, with extreme tenderness to

palpation. Vesicular lesions were appreciated diffusely on the legs, right shoulder, and lateral eyelid (Figure 1). Differential diagnosis included disseminated zoster, drug reaction, Kaposi's sarcoma, Sweet's syndrome, cellulitis, dyshidrotic eczema, or other neutrophilic dermatosis. Given the presence of grouped vesicles, despite a mixed dermatomal and non-dermatomal pattern, our patient was empirically started on intravenous acyclovir 700mg three times daily for herpes zoster.

Punch biopsy over the right anterior clavicle showed a partially necrotic blister containing multi-nucleated cells with nuclear molding and chromatin margination (Figure 2). This was consistent with diagnosis of disseminated



Figure 1: (A) Confluent, violaceous plaque extending from mid-chin to right posterior neck, posterior right scalp, and right clavicle, (B) Confluent, violaceous plaque with vesicles extending to mid-chin, and (C) Diffuse vesicular lesions on left lateral thigh.

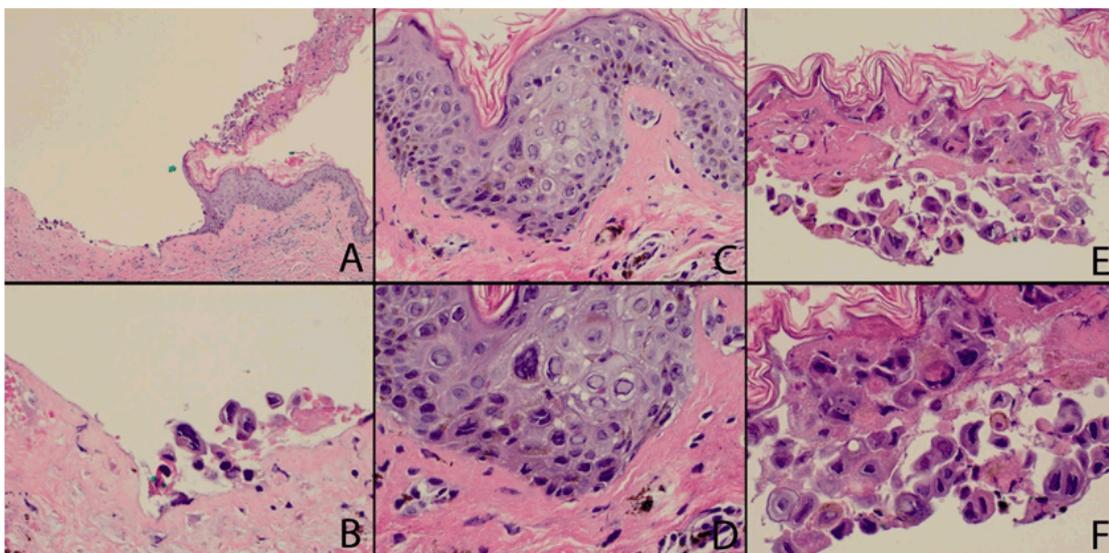


Figure 2: (A) Vesicle (H&E stain, magnification x200), (B) Base of vesicle (H&E stain, magnification x600), (C, D) Intradermal multinucleation, molding, and margination (H&E stain, magnification x400, x600), and (E, F) Roof of vesicle showing acantholysis (H&E stain, magnification x400, x600).

herpes zoster. The absence of eye pain and conjunctival injection excluded the diagnosis of zoster ophthalmicus. The patient continued intravenous acyclovir 700 mg three times daily for a total of 3 weeks, followed by valacyclovir 1 g twice daily for 1 week. Given the patient's immune status, she remained hospitalized for treatment. The patient was hospitalized for a total of 28 days. At first-month follow-up, the rashes had completely resolved.

## DISCUSSION

Atypical generalized zoster is seen almost exclusively in immunocompromised patients. Cases in literature almost exclusively describe patients who are post-renal transplant or with hematogenous malignancy [2, 3]. Our patient presents a unique case of atypical generalized zoster in a solid tumor patient undergoing chemotherapy for stage IV leiomyosarcoma. Our patient showed an atypical presentation with both dermatomal and non-dermatomal plaques in addition to the more typical diffuse vesicular lesions.

Clinical presentations of disseminated VZV can pose diagnostic and therapeutic challenges. Given the rarity of generalized zoster reported in solid malignancy patients, it is unknown whether the presentation or risks of VZV are similar to those with renal transplant or hematogenous malignancy. We assume that the risks would prove comparable, as the mechanism for zoster in both elderly and immunocompromised is a waning of VZV-specific T-cell immunity [4]. As noted, our patient did demonstrate a unique clinical presentation. It is interesting to postulate why the facial and upper extremity lesions were more confluent in our patient, while the remainder exhibited a more generalized distribution—perhaps she was less immunosuppressed than typical post-renal transplant or bone marrow transplant recipients on immunosuppressive agents.

Patients with generalized VZV infection have a high risk of disseminated visceral involvement of up to 50% and mortality of 28–80%—the most frequent causes of death are from pneumonia, encephalitis, and hepatitis, which our patient did not exhibit [5]. Unfortunately, conventional laboratory methods often fail to diagnose VZV in immunocompromised patients—the Tzanck test, electron microscopy, and DIF tend to be less helpful in the setting of atypical zoster. In these cases, PCR may provide a rapid and accurate diagnostic tool with a high degree of specificity [5]. Recent studies have shown that VZV-specific CD4-T cells in patients with zoster bear typical features of anergy. This phenotype is reversible and in the future may also be used as a tool for monitoring VZV reactivations in high-risk patients [6].

High-dose intravenous acyclovir, aggressive supportive care, and zoster immunoglobulin are mainstays of treatment for generalized zoster [5]. Ganciclovir has also shown similar efficacy to acyclovir in VZV, but is limited by its hematologic side effects [7].

## CONCLUSION

With early recognition via clinical evaluation and biopsy, and immediate treatment with acyclovir, our patient was able to achieve complete resolution. We support maintaining a high level of clinical suspicion and a low threshold for treatment of suspected disseminated zoster in immunosuppressed, solid tumor patients. Moreover, the importance of zoster vaccination prior to immunosuppressive therapy is paramount to preventing disseminated cutaneous zoster in this population.

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## Author Contributions

Ashley Nicole Elsensohn – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Maryam Liaqat – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Dante Garcia – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Charay Jennings-Dover – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Gregory Simpson – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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