

Case Scenario 2

A 29 y.o. female commercial sex worker in a large naval port city on the West Coast presents to an emergency room late in the evening of February 12 with fever and an extensive pustular eruption on the face, neck, and anterior elbows. She complains of the onset of a vesicular rash in these areas 36 hours before and increasing symptoms of weakness, fatigue, and malaise. The patient has a fever of 103 F and a rash that looks similar to that seen in the patients pictured on slide 1. This rash is present on the face, neck, and anterior elbows but lesions are not noted elsewhere. Tender cervical and axillary lymphadenopathy is present.

The patient gives a history of an erythematous, itchy, flaky skin eruption in these areas since early childhood that waxes and wanes, recently, the skin rash had been slightly active. The patient does not know of any contact with a known smallpox vaccinee but her clientele includes military personnel as well as civilians. The ER physician calls his state health department to obtain guidance.

1. What is the most likely diagnosis?

Eczema vaccinatum due to contact with a smallpox vaccinee. The diagnosis of EV is supported by the vesiculo-pustular nature of the rash, the regionally extensive but not generalized nature of the rash, and by the history of active skin disease suggestive of atopic dermatitis.

2. What should be done immediately with the patient?

The patient should be hospitalized for supportive care and a consultation with CDC should be consulted to request VIG. EV is associated with a high mortality rate; VIG can significantly reduce this mortality.

3. Should this patient be investigated for other medical conditions for diagnostic or prognostic purposes?

Herpes simplex virus should be excluded as a cause of the rash as herpes can cause an identical rash in patients with atopic dermatitis and the patient is at increased risk for herpes simplex virus infection. Culture, antigen detection, or PCR techniques are likely to be available locally and should be utilized to exclude herpes.

EV was not associated with identifiable immunodeficiencies in the past and an evaluation for immunodeficiency is not routinely recommended for patients with EV. However, this patient is at increased risk for HIV infection and it is reasonable that disorders of cell-mediated immunity may adversely affect the prognosis of EV.

4. What is the usual course of the patient's condition?

The evolution of the individual vaccinia lesions in EV follows the course of normal smallpox vaccination lesions. Patients with extensive EV are frequently severely systemically ill. The clinical picture can resemble that of severe burn patients and these patients require close supportive care with attention to hemodynamic support, secondary infections, and skin care. High doses of IM VIG were frequently used in the past and repeated doses of VIG IV may be needed.

5. If CDC is consulted on this patient, what type of additional contact with CDC will occur?

After VIG is administered to this patient, daily data on the patient's clinical condition and routine laboratory values are required under the IND. In cases where VIG is administered, the CDC clinical team (in conjunction with the state AE coordinators) will followup with the treating physician q 24 -48 hours to assess the need for additional VIG until the patient improves.

6. What type of infection control measures are needed for this patient?

The extensive lesions in EV contain large amounts of vaccinia virus. Nosocomial transmission has not been common but has occurred and is primarily linked to transmission via hands/objects in direct contact with the lesions. Standard precautions should greatly reduce the likelihood of transmission. CDC is working with HICPAC to develop more detailed guidelines for infection control.

7. If this is a smallpox vaccine AE, can this case be linked to exposure to military personnel that departed city A on a naval convoy January 17?

Usually EV in contacts presents 5 – 19 days after exposure. With an onset of ~ February 10, this case is unlikely to be linked to exposure prior to January 17.

8. Is there a spectrum of severity for this smallpox vaccine adverse event?

Data from the U.K. in the 1960s suggests that milder forms of EV occur; a significant proportion of patients reported with the diagnosis of EV from dermatologists were not treated with VIG despite its availability. However, we do not have any data that would help us in firmly identifying which patients are likely to follow a milder course.

9. Would the presentation be different if a adult smallpox vaccinee or smallpox vaccinee adult contact has a history of similar skin rash in childhood but not as an adult?

EV has occurred in both adults and children with a history of atopic dermatitis (or 'eczema') with either inactive or active disease at the time of exposure. Anecdotally, EV may be likely to be milder in individuals who have not had active skin disease since childhood.

Goals:

Clinical picture of EV including mild and severe forms

Therapeutic modalities required

Outcome

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