



Pathology Biology Section - 2012

G124 Disseminated Varicella as a Cause of Sudden or Unexpected Death: A Case Report and Literature Review

Dawn B. Holmes, MD, Adrienne E. Segovia, MD, and James A. Filkins, MD, JD, PhD, Cook County Medical Examiner's Office, 2121 West Harrison Street, Chicago, IL 60612*

After attending this presentation, attendees will have a better awareness of the characteristics of disseminated varicella (chickenpox) infection as a rare cause of sudden or unexpected death.

This presentation will impact the forensic science community by presenting the autopsy findings and circumstances surrounding the death of a 28-year-old female who succumbed to disseminated varicella infection and by comparing her case with others reported in the literature.

Introduction: Varicella zoster virus (VZV) is an infection that commonly occurs in childhood and usually has a relatively benign course in immunocompetent individuals. Mortality from VZV infection in immunocompetent individuals is exceedingly rare. In immunocompromised individuals; however, VZV infection often produces severe manifestations, leading to significantly increased mortality.

Case Report: A 28-year-old African American female with a history of immune thrombocytopenic purpura and sarcoidosis presented to the emergency room with an acute onset of a diffuse vesicular rash throughout her body. Her medications included prednisone, and she reported a history of chickenpox as a child. She was subsequently diagnosed with chickenpox and was treated in the hospital with benadryl and calamine lotion for five days. No antiviral medications were administered. The night of her discharge, she complained of shortness of breath. The following morning she was found unresponsive in her bed at home. At autopsy, she was noted to have numerous macular and papular vesicular lesions in various stages throughout the body, predominantly involving the head/face, neck, torso, back, upper extremities, and proximal lower extremities. The lesions also involved the mucosa of the inner surfaces of the upper and lower lips. Several of the vesicular lesions were crusted. On opening the body, numerous erythematous vesicular lesions were noted on the surface of the lungs, liver, and the mucosal surface of the esophagus, trachea, and epiglottis. Multiple enlarged peribronchial, periaortic, and mesenteric lymph nodes were identified; the largest of these measured 3.0 cm in greatest dimensions, consistent with the patient's known history of sarcoidosis. Histologic investigation uncovered multinucleated giant cells in the enlarged lymph nodes with associated hyalinization. There were also patchy hyaline membranes in the lungs with hemosiderin-laden macrophages, scattered multinucleated giant cells, and pulmonary edema. Mucosal lesions revealed ulceration with marked chronic inflammation extending to the submucosal tissue. The Centers for Disease Control and Prevention confirmed via immunohistochemistry the presence of VZV in various tissues. Toxicologic analyses were negative for ethanol, opiates, and cocaine.

Discussion: Mortality from VZV infection is exceedingly rare. The annual varicella death rate in the United States is 0.4 deaths per million. Previous cases of disseminated varicella mortality have involved unvaccinated individuals who had a primary VZV exposure. These individuals were usually immunocompromised to various degrees, although mortality has been reported in otherwise healthy individuals. Consistent with the majority of the cases reported in the literature, this case report involves an immunocompromised subject; interestingly, however, this was not a case of primary VZV exposure.

Reported VZV complications include pneumonia, disseminated intravascular coagulation, septicemia, encephalitis, acute respiratory distress syndrome, nephritis, myocarditis, and myelopathy. VZV pneumonia is the most common complication with the highest mortality, seen in immunocompromised individuals. Case reports in the literature typically describe an insidious onset of pneumonia with dyspnea, usually developing 1-6 days following the appearance of a vesicular rash. A similar course of events was present in this case report.

This case is reported to demonstrate the clinical course and autopsy findings of disseminated varicella infection. Although an extremely rare entity, it remains a cause of sudden or unexpected death that one may encounter in a forensic setting.

Varicella, Chickenpox, Disseminated