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Recommended Citation

Bhatti, M. A., Manglani, C. B., Khan, M. A. (2004). Fatal case of chickenpox in an adult: a case report. *Journal of Pakistan Medical Association*, 54(2), 104-106.

Available at: https://ecommons.aku.edu/pakistan_fhs_mc_med_intern_med/68

Fatal Case of Chickenpox in an Adult: a case report

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Introduction

Varicella Zoster Virus (VZV) is a double stranded DNA virus of a α -herpes virus family which also include herpes simplex virus type 1 and 2. VZV causes chicken pox (varicella) during primary infection, following which the virus establishes the latency in cells of the dorsal root ganglia. Reactivation of latent virus causes shingles (herpes zoster), a disease predominantly occurring in elderly or immunocompromised patients. National incidence rate of chicken pox in Pakistan is unknown. In United States, before the availability of varicella vaccine, varicella disease was responsible for an estimated 4 million cases, 11,000 hospitalizations, and 100 deaths each year. ² Approximately 90% of cases occur in children and rarely in adults. Fifty five percent of varicella-associated deaths occurred among persons aged greater than or equal to 20 years. ³ We report a fatal case of primary varicella in an immunocompetent adult with severe complications resulting in death. The case highlights the importance of vaccination in children as well as high risk adult population in our country.

Case Report

A 42 year old man presented with one week history of intermittent colicky abdominal pain, vomiting and jaundice. Two days prior to admission he developed vesicular skin rash over face, arms and chest. One day prior to admission he passed hard stools mixed with fresh blood. He was recently found to be positive for hepatitis B surface antigen. Four days prior to his illness; two of his children had a febrile illness with vesicular skin rash diagnosed as chickenpox. He was a fisherman by profession, occasionally drank small amount of alcohol and smoked 15 cigarettes daily for the last 30 years. On physical examination, he was jaundiced, had pedal edema and vesicular eruption all over the body particularly in axilla, chest and face (Figure 1). Few of the vesicles were umbilicated. He had conjunctival hemorrhage in both eyes. No lymphadenopathy was detected. Bilateral inspiratory crackles were heard at lung bases. Abdominal, cardiovascular and neurological examinations were unremarkable. Initial laboratory workup showed hemoglobin of 13.8 grams/dl, white blood cells 5.7/cmm and platelets were clumped, creatinine 0.9 mg/dl, total bilirubin 2 mg/dl, serum alanine aminotransferase 213, LDH was 2676 and lipase was 537. [(0)] CT scan of abdomen revealed nodularity in liver and mild ascites. There was moderate splenomegaly and varices noted along the splenic vein. Findings were suggestive of chronic liver disease with portal hypertension. Bilateral soft tissue infiltrates were noted in the lungs consistent with pneumonitis (Figure 2). A Tzanck Smear of vesicular lesion from chest revealed inclusion bodies (Figure 3). Following admission, treatment was started with high dose intravenous acyclovir and meropenam. He required intubation the following day and his clinical condition continued to deteriorate with development of acute renal failure and disseminated intravascular

coagulation. He started bleeding from the mouth, nose and endotracheal tube and did not respond to aggressive management and died on the 4th day of admission.

Discussion

Humans are infected with VZV by contact with respiratory mucosa or conjunctiva. The virus disseminates through the blood stream inside mononuclear cells and to the skin causing the generalized vesicular skin rash of varicella. 4 Primary varicella infection (chicken pox) is a common, highly contagious, vaccine preventable disease. Varicella is usually a self limited disease that lasts 4-5 days and is characterized by fever, malaise, and generalized vesicular rash typically consisting of 250-500 lesions.² The average incubation period for varicella is 14 days and almost all cases occur 10-20 days after exposure. 5 In contrast, reactivation disease (herpes zoster) in immunocompetent individuals is usually a localized illness, affecting the skin innervated by one or two adjacent dermatomes. However, in patients with impaired cell mediated immunity disseminated disease with widespread cutaneous and visceral involvement may develop⁶ and may take the one of four forms: local (classic) zoster, atypical generalized zoster with or without visceral involvement, and visceral zoster without skin lesions. 7 Our case did not have previous history of chicken pox and had no obvious underlying cause for cellular immunodeficiency disorders. His history and diagnostic work-up is suggestive of primary varicella infection (chicken pox). Anecdotal observations and case reports have previously noted that varicella occurs more frequently in adults in tropical than in temperate climates. 8 In general, adults with varicella are more severely ill than children and have a greater incidence of varicella pneumonia and death. 5 Previous analyses of age specific risks have estimated that the risk of death is 25 times more and the risk of developing encephalitis is eight times higher in adults than in children.⁸ In several series of hospitalized adults, primary varicella pneumonia has been observed in more than 14% of patients,⁸ In our country, chicken pox in adults is suspected on clinical grounds and confirmed either by tzanck smear or type specific serology. VZV can be recovered on culture of the vesicular fluid for a few days after onset of rash but specific laboratory facility for isolation of the virus are not commonly available in our country. Varicella (chicken pox) in adolescents and adults tends to be more severe, and should be treated promptly, preferably within 24 hours of the onset of rash. 5 IV acyclovir is the drug of choice. Newer formulation like valacyclovir or famciclovir could be used orally. Varicella can be prevented in a recently exposed unvaccinated or immunocompromised person by the administration of varicella-zoster immune globulin. It must be given within 96 hours of exposure to be effective. 9 Our patient did not survive even with aggressive management and manifested multiple complications including pneumonitis, pancreatitis and disseminated intravascular coagulation. The reason for poor response was probably the delayed presentation to hospital which is a common problem in managing these cases in our country. This case highlights the importance of increasing public awareness, vaccination, early recognition and diagnosis by community physicians, and prompt treatment or referral to the hospital for management to prevent fatal complications. The vaccination of all children and susceptible adults should be considered and may decrease the incidence of this preventable disease.

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