Case Report

Progressive varicella syndrome in the setting of pediatric AIDS: An eye opener

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Abstract

Varicella zoster virus (VZV) infections are known to be atypical and severe in immunocompromised patients. An eight-year-old girl presented with extremely painful, atypical skin lesions and features of meningitis and pneumonitis. On investigation, she was found to be human immunodeficiency virus (HIV) infected, with very low CD4 count. A diagnosis of ‘progressive varicella syndrome’ was made, and the child was started on antiretroviral therapy and IV acyclovir. This resulted in a complete resolution of all the clinical features. However, the skin lesions promptly relapsed when acyclovir was withdrawn. Oral Acyclovir was started, and had to be continued to keep the disease under control.

Key words: Acyclovir, AIDS, progressive varicella syndrome

INTRODUCTION

Children with AIDS are more likely to develop disseminated and atypical varicella zoster virus (VZV) infections. Newer varicella syndromes have been described in pediatric HIV. These patients may develop Herpes Zoster soon after chicken pox, or may develop ‘recurrent varicella syndrome’ or ‘progressive varicella syndrome’.

Recurrent varicella syndrome is associated with a modest decrease in CD4 count. Here, a new episode of disseminated skin lesions occurs at least one month after a chicken pox attack, in the absence of exposure. Progressive varicella syndrome is associated with very low CD4 count, and is characterized by new skin lesions which continue to appear for at least one month duration.

CASE REPORT

An eight-year-old girl presented with painful blisters on the skin of 40 days duration. She also had fever with chills, cough, dyspnoea, and severe headache for the past one month. She had suffered from chickenpox four months back, which resolved uneventfully. She was anemic, and had stunted growth. Chest examination revealed bilateral crepitations. She also had neck stiffness.

Multiple painful and tender vesicles and bullae of varying size were seen all over the body, predominantly over the acral areas. Few bullae were hemorrhagic. Warty excrescences were seen overlying some blisters. Oral erosions were noted. The skin lesions started as tiny vesicles [Figure 1], which evolved into larger bullae, some of them were hemorrhagic [Figure 2]. Thereafter, they progressed to form warty excrescences [Figure 3], and finally resolved with varioliform scars. New lesions continued to appear, even as the older ones healed. Oral erosions were also present. There was no dermatomal grouping of the skin lesions.

Due to the unusual morphology of the lesions, enzyme linked immunosorbent assay (ELISA) for HIV was done, which turned out to be positive. Western blot for HIV was also positive. Her parents were HIV negative. There was no history suggestive of sexual abuse. No possible explanation could be offered for the HIV positivity of the child, other than frequent,
and possibly unsafe intramuscular injections in the past.

Tzanck smear from the blisters showed multinucleated giant cells [Figure 4]. Histopathology of the early vesicle revealed typical features of varicella blisters, and that of late warty lesions showed hyperkeratosis, acanthosis, and papillomatosis without the classical features of viral blister.

Chest x-ray showed pneumonitis, cerebrospinal fluid (CSF) analysis revealed lymphocytosis. Complete hemogram was normal except for dimorphic anemia. Her CD4 count was 41, blood VDRL was nonreactive, and serum IgG and IgM for HSV-1 and HSV-2 were negative. Serum IgM for VZV was negative, but IgG for VZV was positive in very high titre.

The patient was started on antiretroviral therapy comprising three drugs: stavudine, lamivudine, and nevirapine. Intravenous acyclovir in the dose of 10 mg/kg body weight was administered for three weeks. There was dramatic improvement with complete resolution of the skin lesions, meningitis, and pneumonitis. At the end of third week, acyclovir was stopped and the patient was discharged. However, the blisters promptly recurred within two weeks. This time oral acyclovir was started with the dose of 80 mg/kg/day in four divided doses, and the lesions subsided again. At present, the child is on antiretroviral therapy and oral acyclovir with the dose of 80 mg/kg/day in four divided doses, for the past three months. Her general condition is improving, but the occasional vesicle still continues to appear. It has been planned to continue acyclovir with the same dose till her CD4 count improves substantially.

DISCUSSION

Progressive varicella syndrome has been documented in children with leukemia,[2] Wiskott-Aldrich Syndrome[3] and advanced HIV infection, it occurs...
when the CD4 count is very low[1] and is associated with internal organ involvement such as meningitis, and pneumonitis, which can be fatal. By definition, the skin lesions continue to appear for at least one month. Intravenous foscarnet may be needed for cases that do not respond to acyclovir.

Verrucous lesions have been described in Herpes Zoster in the setting of AIDS. Our case assumes greater significance due to the presence of prominent warty lesions in progressive varicella syndrome, which, to the best of our knowledge, has not been reported so far.

The pathogenesis of this condition is thought to be due to altered gene expression of VZV, leading to reduced synthesis of major late envelope glycoproteins gE and gB, resulting in more indolent infection.[4]

The significance of the genetic evolution of VZV and its clinical relevance, in the context of HIV pandemic, merits further attention. Questions that are yet to be answered include the drug of choice in the new varicella syndromes and the appropriate dosage regimen. The CD4 count at which antivaricella treatment can be safely withdrawn without the risk of relapse also needs to be elucidated.

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