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PROGRESSIVE VARICELLA SYNDROME WITH *VARICELLA GANGRENOSA* IN AN IMMUNE-COMPETENT INFANT

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Abstract
Varicella is common and highly contagious and affects nearly all susceptible children before adolescence. Progressive varicella syndrome is a severe complication of primary Varicella Zoster Virus (VZV) infection, with visceral organ involvement, coagulopathy, severe hemorrhage, and continued vesicular lesion development. We report a rare case of progressive varicella syndrome with varicella gangrenosa in a previously well female child of ten months. She presented with history of recurrent vesiculo-bullous skin lesions involving the chest, back and extremities since two months with dry gangrene of 1st, 3rd and right great toe. VZV Polymerase Chain Reaction (PCR) of vesicle fluid was positive. Workup for immunodeficiency state was negative. She responded dramatically to intravenous acyclovir.

Key words: progressive varicella syndrome; varicella gangrenosa; immune-competent

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Introduction

Varicella, commonly known as chickenpox, is caused by the varicella-zoster virus and causes primary, latent, and recurrent infections. Varicella is common and highly contagious and affects nearly all susceptible children before adolescence. Infections with varicella-zoster virus (VZV) are usually considered benign infections. However, severe complications including bacterial super infections, coagulopathies, and central nervous system manifestations with a potentially fatal or long term disabling outcome can occur [1,2].

Although most varicella infection confers life-long immunity, clinical reinfections among healthy children have been described [3]. Progressive varicella is a severe complication of primary VZV infection, with visceral organ involvement, coagulopathy, severe hemorrhage, and continued vesicular lesion development [4]. Gangrene of skin and deeper tissues is an unusual complication of varicella. The term varicella gangrenosa has been applied such conditions. However, varicella gangrenosa is a rare complication of this disease, infrequently reported in the literature [5,6].

Very few cases of progressive varicella syndrome have been reported in literature that too in immuno-compromised host. We report a case of progressive varicella syndrome with varicella gangrenosa in an immune-competent female child of ten months.

Case report

A ten month old female baby to us with history of recurrent vesiculo-bullous skin lesions involving the chest, back and extremities since two months with recent progression to palms and perianal area, abdominal distention, tachypnoea, swelling of bilateral lower limbs and discoloration of toes of right foot.

There was no previous history of recurrent infections or recurrent skin lesions; however she had history of vesiculo-bullous lesions in elder sibling four month back. The child was admitted three times elsewhere and treated with multiple antibiotics with inadequate response.

On admission child was lethargic, had multiple confluent hemorrhagic, vesiculo-bullous lesions all over body (Fig. 1), anasarca predominantly in bilateral lower limb with dry gangrene of 1st,3rd and great toe (right) and bilateral crepitations in chest. Investigations revealed normal complete blood count, normal liver functions with low serum albumin (2g/dl) and normal coagulation profile. Blood culture was sterile. VZV Polymerase Chain Reaction (PCR) of vesicle fluid was positive. Chest x-ray was suggestive of bronchopneumonia and Doppler of bilateral lower limbs was normal. Her immunoglobulin levels were normal, Nitro Blue Tetrazolium test (NBT) was normal and P24 antigen assay for HIV was negative.

Patient responded dramatically to intravenous acyclovir. Low molecular weight dextran, Low molecular weight heparin and Pentoxifylline were administered for gangrene. Her general condition improved and her lesions started healing by day five and she was discharged after fifteen days.

Figure 1. A. Vesiculo-bullous lesions with ulceration over whole back. B. Vesiculo-bullous lesions over thigh, chest and abdomen. C. Edematous lower limbs with lesions. D. Confluenced vesiculo-bullous lesions.

Discussion

Progressive varicella, with visceral organ involvement, coagulopathy, severe hemorrhage, and continued vesicular lesion development, is a severe complication of primary VZV infection [4]. Varicella gangrenosa is a very rare complication [5,6]. Our patient presented with features of progressive varicella along with dry gangrene of toes. Although rare in healthy children, the risk for progressive varicella is highest in children with congenital cellular immune deficiency disorders and those with malignancy [4]. Progressive varicella syndrome has been documented in children with leukemia [7], Wiskott-Aldrich Syndrome [8] and advanced HIV infection, it occurs when the CD4 count is very low [9] 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. Our patient had skin lesions for two months and was an immune-competent infant. Intravenous foscarnet may be needed for cases that do not respond to acyclovir, however our patient responded well to intravenous acyclovir.

Conclusion

Our encounter with this case highlights that although rare progressive varicella can present in immune-competent child. Prompt diagnosis and treatment with acyclovir leads to complete recovery.

Authors' contributions

SB, NB, KU and MJ were involved in patient management; SB, KU and MJ were involved in manuscript preparation; SB, NB were involved in reviewing the manuscript and final approval.

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