

Extensive Varioliform and Atypical Targetoid Presentation of Acute Varicella in a Patient of Ankylosing Spondylitis

Dear Sir,

Majority of acute varicella episodes are mild and self-limiting, and cutaneous features include vesicles and bullae over erythematous background. However, in the immunocompromised, the rash maybe extensive and hemorrhagic with systemic involvement.^[1] We hereby report a young patient of ankylosing spondylitis with atypical presentation of extensive varicella.

An 18-year-old male with ankylosing spondylitis, (HLA-B27 positive) previously on treatment with oral methotrexate (15 mg/week) and deflazacort (18 mg BD), presented with complaints of high-grade fever with multiple vesicles and crusted lesions all over the body since 5 days. The vesicular eruption started over forehead and gradually involved entire body within 2 days. His parents denied any childhood history of either varicella infection or immunization. Treatment with oral valacyclovir (1 g TDS for 5 days) provided only 10–20% relief in lesions and new lesions continued to appear. Patient was admitted and examination revealed vesicles and predominantly tense, turbid fluid-filled bullae, on background of normal skin. Many of the lesions on trunk and proximal extremities showed central depression [Figure 1a]. Lower limb and back lesions prominently showed atypical targetoid morphology [Figure 1b] and on back, skin at periphery of the bullae was peeling with tangential pull [Figure 1c]. Lesions were more confluent over face and trunk than limbs. Additionally, there were crusted ulcers over face (forehead), hard palate, back, and glans. Tzanck smear revealed multinucleate giant cells and histopathology revealed intra-epidermal split and reticular degeneration. Routine blood investigations including viral markers (hepatitis B and C and HIV) and chest radiology were normal. Patient was diagnosed with extensive varicella and treated with intravenous acyclovir (10 mg/kg/dose TDS

for 7 days, followed by oral acyclovir (800 mg 5 times a day for 1 day) and all lesions healed with scab formation. On follow-up [Figure 2a and b], patient was most disturbed by atrophic scars especially over the head and neck region.

In acute varicella, occasionally the virus may disseminate viscerally and viral DNA maybe detectable in several organs using PCR. In immunocompetent adults, the most common presentation of dissemination is pneumonia, though hepatitis, meningoencephalitis, and renal failure have also been described.^[2] The immunocompromised are at a greater risk of pneumonitis and visceral involvement and may experience invasive disease, culminating in multi-organ failure.^[3]

Disfiguring pock-like facial scarring was among the known sequelae of variola major (small pox). However, the majority of varicella lesions heal with no residual changes or only post-inflammatory hyperpigmentation on facial skin. Around 18% develop atrophic scarring following the disease and less than 2.5% of patients have five or more such scars.^[4] The peculiar findings in our patient included widespread tense and turbid vesicles in the absence of background erythema, atypical targetoid morphology, and severe scarring, even involving glans penis. The easy peeling of skin over back was akin to pseudo-nikolsky sign. Eventhough exact reason is unknown and similar finding doesn't find mention in English language literature, it could possibly be explained by excessive disease activity. Therefore, a low threshold should be kept for diagnosing acute varicella in immunocompromised patients and prolonged/parenteral antiviral therapy may be considered in view of poor initial response.

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Figure 1: (a) Multiple tense and turbid vesicles over trunk and proximal extremities with central depression. (b) Multiple vesicles over bilateral thighs and scrotum with dusky, atypical targetoid morphology. (c) Skin peeled off at periphery of the bullae due to tangential pressure (Pseudo-Nikolsky sign)



Figure 2: Multiple hypopigmented atrophic scars involving a) Peri-auricular region with involvement of side of neck (at 18 months' follow-up) and (b) Glans penis (at 2 months' follow-up)

Conflicts of interest

There are no conflicts of interest.

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