Original Research Paper

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Paediatrics

A RARE VARICELLA ZOSTER INFECTION COMPLICATED BY PURPURA FULMINANS IN A CHILD.

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ABSTRACT Varicella is a viral exanthematous illness caused by the varicella-zoster virus. Usually, it is mild and self-limiting, but severe or life-threatening complications can occur. Purpura fulminans, being one of them, is a potentially life-threatening condition of intravascular thrombosis and hemorrhagic infarction of the skin. We present the case of a 7-year-old girl with post varicella purpura fulminans leading to chronic non-healing ulcers on bilateral dorsum of the feet. Despite having normal laboratory results initially, the condition was promptly diagnosed and treated with antibiotics as per sensitivity pattern and wound debridement.

KEYWORDS : Purpura fulminans, Post varicella, Varicella complications, chronic non-healing ulcers.

INTRODUCTION:

Varicella-associated purpura fulminans (PF) is a rare syndrome associated with substantial morbidity and mortality. It is characteristically associated with autoimmune protein S deficiency and profound hypofibrinogenemia[1].

Purpura fulminans is used to describe a heterogeneous group of disorders characterized by rapidly progressive purpuric lesions, which may develop into extensive areas of skin necrosis and peripheral gangrene typically distributed symmetrically on the lower extremities and buttocks. Laboratory evidence of consumptive coagulopathy is seen in this disorder. The histopathologic findings are extensive thrombosis of the dermal capillaries and venules with haemorrhagic infarction of the surrounding tissues[2].

We present a case of such a child, whose normal lab results at presentation made the diagnosis difficult.

Case Report:

A 7-year-old female child, with height and weight for age within median to -1SD, not immunised for Varicella, presented with bilateral chronic ulcers over dorsum of the feet for 2 months. Two months ago, the child had fever with fluid-filled vesicular rash diagnosed as varicella. She received symptomatic treatment for varicella. After 5 days she developed swelling over both lower limbs for which she was hospitalized for 3 days. Ten days later there was blackish discolouration of both feet; hence child was referred to our centre and was admitted.

On examination, the child was afebrile, HR: 100/min, RR: 26/min, BP: 104/60 mm of Hg, SpO2: 98%. Mild pallor+, post varicella scar marks were seen. Chronic non-healing ulcers with thick margins were present on bilateral dorsum of the feet approximately 10 x 6 cms. (Fig 1a and 1b). Systemic examination was within normal limits.

Investigations:

There was mild anemia, Hb: 10.3gm/dl, leucocytosis 23.6 X 10^3/ml, thrombocytosis 1260 x10^3/ml. CRP was Positive, fibrinogen was elevated (614.5 mg/dl), with raised D-dimer (3566.14 ng/ml). Prothrombin time, INR, Protein C, Protein S, and Anti-thrombin III were within normal limits. HIV seronegative, Covid Antibody was negative. Ultrasound of lower limbs s/o diffuse subcutaneous oedema with normal B/L lower limb doppler. Wound swab culture sensitivity- Growth of Pseudomonas aeruginosa. (Table 1).

Table 1

Laboratory Test	Result	Reference Range
Hb (Hemoglobin)	10.3 gm/dl	11.5-13.5 gm/dl
TLC (Total Leukocyte Count)	23.6 X 103/ml	4.5-13.5 X 103/ml
Platelets	1260 X 103/ml	150-450 X103/ml
CRP (C-reactive protein)	77.86 (Positive)	-
PT (Prothrombin Time)	14.6 seconds	11.7-15.1 seconds
APTT (Activated Partial Thromboplastin Time)	25.1 seconds	31.8-43.7 seconds
INR (International Normalized Ratio)	1.13	0.87-1.20
Fibrinogen	614.5 mg/dl	200-400 mg/dl
D-dimer	3566.14 ng/ml	100-560 ng/ml
Protein C activity	99%	70-140%
Free Protein S level	120.9%	54.7-123.7%
Varicella Serum IgM	Positive (1.533)	>= 1 positive
Anti Thrombin-III	111%	95-134%
HIV	Negative	-
USG lower limbs B/L lower limb doppler	Diffuse subcutaneous oedema Normal	-
Covid IgG Antibody	Negative	-
Wound swab culture sensitivity	Growth of Pseudomonas aeruginosa.	Sensitive to Piperacillin-Tazobactam, Amikacin, Gentamicin, Cefepime, Aztreonam.
Blood culture sensitivity	No growth detected	-

Treatment Received:

IV Piperacillin-tazobactam and Gentamicin was given as per culture sensitivity for 14 days. The repeat wound swab was sterile. Debridement was done by the Plastic surgery unit. Daily dressing with Tulle Gras impregnated with Chlorhexidine acetate, paraffin, Papain, and Urea ointment (Debridement agents) was done.

Outcome:

Ulcers eventually healed and raw areas were covered naturally after 4 weeks. (Fig2a and Fig2b)

Skin Grafting was not required.

DISCUSSION:

PF is a clinical diagnosis requiring a high index of suspicion early in the patient's presentation. [4] It is an acute, rapidly developing haemorrhagic skin necrosis due to dermal vascular necrosis associated with disseminated intravascular coagulation. Postinfectious PF usually occurs 7 to 10 days after the onset of symptoms of acute varicella infection. [2] The ecchymotic lesions are typically distributed symmetrically on the lower extremities and buttocks.

Transient deficiency of protein S activity is considered as a possibility in the development of varicella-associated PF due to the induction of anti-protein S autoantibodies. These antibodies remain for a few months, and protein S activity returns to normal.

In contrast to previously reported cases, our patient's protein C and protein S levels were normal, which, could be because of the late presentation to our hospital, i.e, 2 months after varicella infection.

Fibrinogen can be low, normal, or high. Infection tends to increase fibrinogen, whereas DIC may consume fibrinogen. However, a normal fibrinogen level excludes neither DIC nor purpura fulminans.[3]

Because PF is primarily a thrombotic process, prompt heparinization combined with aggressive blood product replacement using cryoprecipitates, protein C, and antithrombin concentrates to replace the factors consumed, is indicated. In conjunction with steroid therapy, plasma exchange reduces the titre of the protein S autoantibodies. If progressive life- or limb-threatening thrombosis occurs despite these measures, then fibrinolytic therapy should be considered. Anticoagulation therapy should be continued until the free protein S levels return to normal.[5]

In few extreme cases, amputation of limbs has been required.

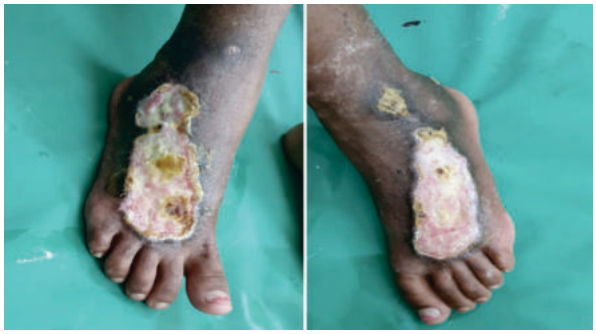

Fig 1a: Right foot- chronic non-healing ulcer.
Fig 1b: Left foot-chronic non-healing ulcer.

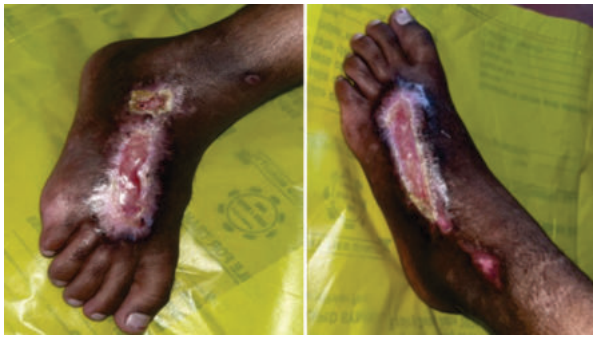

Fig 2a: Right foot- healing ulcer with healthy margins and new skin cover after 4 weeks
Fig 2b: Left foot- healing ulcer with healthy margins with new skin cover after 4 weeks

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