A Fatal Case of Febrile Ulceronecrotic Mucha–Habermann Disease in a 10-Year-Old Boy

Dear editor,

Febrile ulceronecrotic Mucha–Habermann disease (FUMHD), also known as pityriasis lichenoides et varioliformis acuta (PLEVA) fulminans, is a rare, severe variant of PLEVA, characterized by crops of rapidly developing hemorrhagic vesicles, bullae, crusted papules, and plaques progressing to innumerable, coalescent, ulceronecrotic lesions along with fever and systemic features and having high mortality.[1] The exact pathogenesis of FUMHD is still obscure, and there are no laid-out treatment guidelines due to the rarity of the condition. Systemic high-dose steroid, intravenous immunoglobulin (IVIG), methotrexate, cyclosporine, cyclophosphamide, phototherapy, acyclovir, dapsone, pentoxyphylline, tumor necrosis factor-alpha inhibitor, and skin grafting have been tried so far with inconsistent results.[1] Since this condition was first defined by Degos e*t al.*,[2] a total of 70 cases with 11 deaths have been reported so far. Children appear to have better outcome, as there is only a single published case of mortality in pediatric age.

Our patient, a 10-year-old boy, presented with history of progressive pleomorphic lesions (papules, plaques, vesicles, erosions with hemorrhagic crusts) for 3 weeks duration [Figure 1a, b], followed by exacerbation of lesions in the form of coalescing ulcers and erosions over the last 1 week after application of neem (*Azadirachta indica*) and turmeric paste. There was no history of any fever, upper respiratory tract infection, diarrhea, and drug intake prior to onset of these lesions. At the time of admission in Dermatology ICU, he was febrile (99.8°F) and weighed 34 kg; dermatological examinations revealed widespread erosions covered with hemorrhagic crusts smeared with paste, hemorrhagic vesicles, predominantly on trunk, axillae, groin folds, and proximal extremities associated with facial and scrotal edema covering almost 50% of the body surface area (BSA) [Figure 2a, b], sparing part of face, acral areas of extremities, oral and genital mucosa. Nikolsky's sign was negative.

His baseline and follow-up investigations are presented in Table 1. Histopathologic examination revealed features consistent with PLEVA [Figure 3a, b]. The child was started on injection piperacillin–tazobactam in combination with teicoplanin, intravenous daily methyl prednisolone at a dose of 15 mg/kg/d for 3 days, IVIG 2 g/kg over a period of 3 days followed by tab prednisolone 0.5 mg/kg/d. Supportive measures like air-fluidized bed, ambient temperature of 30°C, nanocrystalline silver dressing (Acticoat®), potassium permanganate soaks, care of wound, fluid electrolyte balance, and high calorie and protein diet were ensured, while maintaining strict reverse barrier nursing as per burn patient protocol. Even after administering injection methotrexate, dexamethasone, plus oral cyclosporine, low-molecular-weight heparin (LMWH), and upgradation of antibiotic and antifungal through meropenem, tigecycline, colistin, and caspofungin, he continued to have ulceronecrotic lesions along with intermittent high-grade fever [Figure 4a, b]. Despite consistent and co-ordinated efforts from a multispeciality group, regular revision of antibiotics based on blood and pus cultures, and administration of intravenous albumin, packed RBC, and granulocyte colony-stimulating factor (G-CSF), the child's condition continued to deteriorate and he developed growth of *Pseudomonas aeruginosa* on the skin ulcers and blood culture, followed by features of severe sepsis and acute respiratory distress syndrome (ARDS) and finally succumbed to his illness on day 34 of hospitalization.

Clinically as well histopathologically, he was a classic case of PLEVA going to FUMHD according to the criteria set up by Nofal *et al*. for the diagnosis of FUMHD.[1] Erythema multiforme, hemorrhagic chickenpox, lymphoid papulosis, hemorrhagic pityriasis rosea, guttate psoriasis, lichen planus, Gianotti–Crosti syndrome, and Stevens–Johnson syndrome (SJS) can mimic FUMHD at times.[1] The etiology of FUMHD is still elusive, but hypersensitivity reaction to multiple viral agents, for example, herpes simplex virus, parvovirus B19, varicella zoster virus, Epstein–Barr virus, cytomegalovirus, adenovirus, measles virus, and HIV, has been implicated as a cause. So far, only eight cases (12.5%) have shown possible link to underlying infective origin.[1] Some studies have suggested that the highly aggressive course or mortality in FUMHD may be due to presence of monoclonal T cells, which may be a variant of cutaneous T-cell lymphoma,[3] while others have refuted this association.[4] Being a very rare disease, there is inconsistency and scarcity of literature on its therapeutics. There are few encouraging reports of methotrexate, but the numbers are very limited and most of these cases did not have severe FUMHD[5,6] unlike our patient. We gave preference to methyl prednisolone pulse, thinking about extensive irritant contact dermatitis following application of neem and turmeric paste. We covered the possible infective agents and combined IVIG along with systemic steroid for better efficacy. He did show mild improvement initially, but subsequently, he continued to develop ulceronecrotic lesions. Later, we added methotrexate and cyclosporine, but nothing worked in this case. His admission to tertiary care was delayed by a week. He had very extensive, deep ulceronecrotic lesions that were prone to secondary

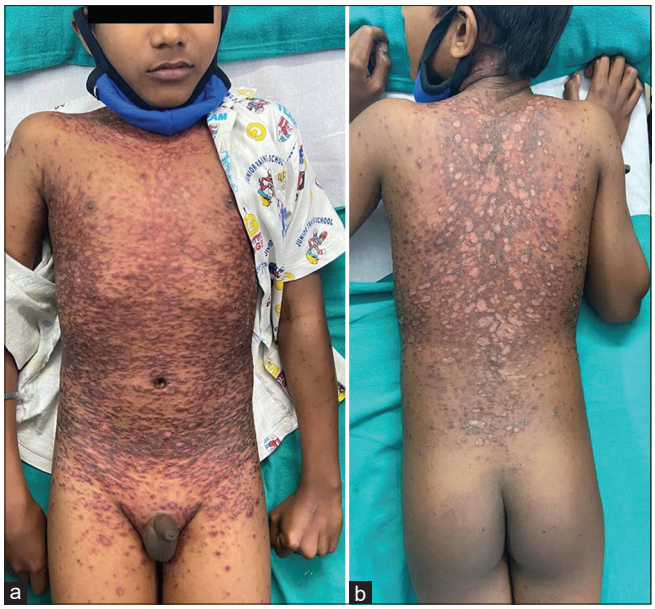

Figure 1: (a) Discrete as well as coalescing papules, plaques, hemorrhagic vesicles, and erosions covered with hemorrhagic crusts present over the front of trunk. (b) Discrete as well as coalescing papules, plaques, hemorrhagic vesicles, and erosions covered with hemorrhagic crusts present over the back of trunk

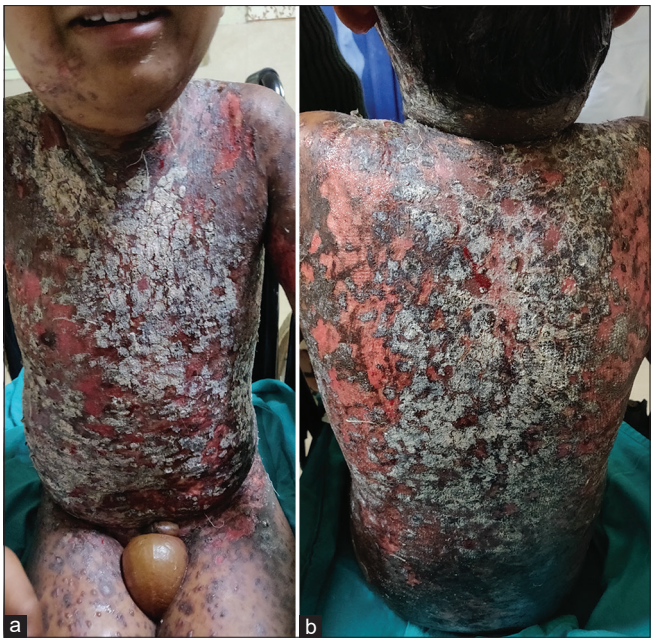

Figure 2: (a) Widespread erosions covered with hemorrhagic crusts, smeared with ayurvedic paste on the front of trunk; discrete and coalescing hemorrhagic vesicles, bullae, erosions, purpuric macules on the extremities; groin with edema of scrotum. (b) Widespread erosions covered with hemorrhagic crusts, smeared with ayurvedic paste on the back

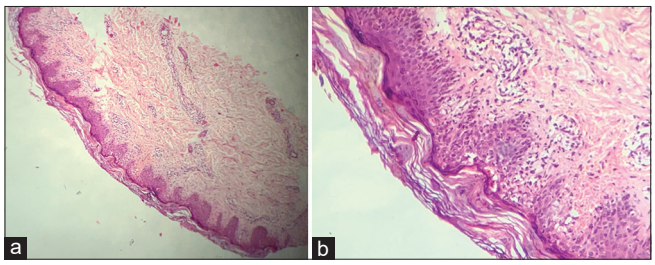

Figure 3: (a) Histopathology (20×): H and E–stained section shows tissue lined by keratinized stratified squamous epithelium. Epidermis shows parakeratosis and focal spongiosis, while mononuclear inflammatory infiltrate is present in the dermis. (b) Histopathology (40×) reveals basal cell vacuolation and few necrotic keratinocytes with apoptotic bodies in lower epidermis. There is mild mononuclear inflammatory infiltrate with extravasation of RBC in the dermis

Figure 4: (a) Progression of skin lesions on the front of trunk and extremities with ulceronecrotic lesions, fresh hemorrhagic vesicles, blackish-greenish slough, purpuric macules on the extremities. Few areas on the upper extremities show minimal resolution of pre-existing erosions. (b) Progression of skin lesions on the back with deeper ulceronecrotic lesions and blackish-greenish slough

infection. On the hindsight, we thought widespread debridement of necrotic tissues under general anesthesia, venesection at a healthy area prior to placing central line, strict vigilance of barrier nursing, and nasogastric feed could have altered the outcome of the patient. The dermatologists should be encouraged to publish as many cases of PLEVA and FUMHD for a wide understanding of the disease along with its treatment modalities.

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Table 1: Results of the investigations conducted over the period of hospitalization

Laboratory parameters		At the time of admission	First week	Second week	Third week	Fourth week	Fifth week
Complete blood count and blood sugar	Hb%	12.4	11.2	9.2	8.5	6.5	7.3
	Total leukocyte count (cells/mm^3) with differential leukocyte count (PLME) and platelet count (lakh/mL)	7800 P70L26M1E03 2.34	8300 P64L33M2E01 2.35	3800 P57L37M1E05 1.14	2400 P70L27M1E2 1.12	2200 P63L34M1E02 1.04	1800 P60L37M1E02 0.92
Blood sugar	Blood sugar (random) (mg/dL)	128	130	136	138	128	110
Renal function test	Urea/serum creatinine (mg/dL)	41/0.48	43/0.7	51/0.8	56/1.1	56/1.2	52/1.4
Liver function test	Serum bilirubin (mg/dL)	0.6	0.7	0.8	1.2	1.6	1.8
	Total protein (g/dL)	4.8	4.2	4	4.2	4	3.6
	Albumin (g/dL)	2.7	2.5	1.6	1.8	1.6	1.5
	SGOT (U/L)	42	76	85	108	115	130
	SGPT (U/L)	34	82	122	175	180	221
Serum electrolytes	Na/K (mmol/L)	147/4.4	138/4.7	139/3.2	139/3.4	141/4.1	142/3.5
Inflammatory markers	C-reactive protein (mg/dL) Normal <10	14	36	76	140	80	75
	D-dimer (ng/mL) Normal <500	550	640	24000	2700	970	1100
	Serum ferritin (ng/mL) Normal 70-435	530	848	975	1250	1150	1400
	LDH (u/L) Normal 140-280	210	220	300	320	350	380
	Procalcitonin (ng/mL) Normal <0.09	0.02	0.04	0.06	0.8	3	11
Serology of viral and etiological agents	HBV, HCV Ab, HIV, TORCH, ASO titre	Not positive	Not done	Not done	Not done	Not done	Not done
Skin swab culture		No growth	*Acinetobacter baumannii* Sensitive to tigecycline, colistin	*A. baumannii* Sensitive to tigecycline	*A. baumannii* Sensitive to tigecycline, colistin	*A. baumannii* Sensitive to tigecycline, colistin	*Pseudomonas aeruginosa* Sensitive to colistin
Urine examination and culture		No growth	No growth	No growth	No growth	Growth of *Escherichia coli* sensitive to meropenem	No growth
Blood culture		No growth	No growth	No growth	No growth	Growth of *P. aeruginosa* sensitive to colistin and merpenem	Growth of *P. aeruginosa* resistant to all
NAD: No abnormality detected		NAD	Not done	NAD	NAD	NAD	Features of acute respiratory syndrome ARDS
Chest X-ray PA view							

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Conflicts of interest

There are no conflicts of interest.

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References

1. Nofal A, Assaf M, Alakad R, Amer H, Nofal E, Yosef A. Febrile ulceronecrotic Mucha-Habermann disease: Proposed diagnostic criteria and therapeutic evaluation. Int J Dermatol 2016;55:729-38.
2. Nofal A, Alakad R, Assaf M, Nofal E. A fatal case of febrile ulceronecrotic Mucha-Habermann disease in a child. JAAD Case Rep 2016;2:181-5.
3. Cozzio A, Hafner J, Kempf W, Häffner A, Palmedo G, Michaelis S, *et al*. Febrile ulceronecrotic Mucha-Habermann disease with clonality: A cutaneous T-cell lymphoma entity? J Am Acad Dermatol 2004;51:1014-7.
4. Lopez-Estebaranz JL, Vanaclocha F, Gil R, García B, Iglesias L. Febrile ulceronecrotic Mucha-Habermann disease. J Am Acad Dermatol 1993;29:903-6.
5. Lin CY, Cook J, Purvis D. Febrile ulceronecrotic Mucha-Haberman disease: A case with systemic symptoms managed with subcutaneous methotrexate. Australas J Dermatol 2012;53:83-6.
6. Ngo T, Hossain C, Cohen J, McLellan B, Blasiak R, Balagula E. A diagnostically challenging case of febrile ulceronecrotic Mucha-Habermann disease in an adult female successfully treated with methotrexate and cyclosporine. Case Rep Dermatol 2021;13:12-7.

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