A febrile 10-year-old girl was referred for evaluation of a diffuse febrile papulovesicular eruption present for 5 days consisting of rare sparse vesicles with a tiny erythematous halo and some papules on the trunk and legs with mild itching (Figure 1 and Figure 2). Her temperature had fluctuated between 38°C and 40°C, and on the third day, the family pediatrician began acyclovir treatment (800 mg, 5 times daily) for suspected varicella infection, which she had had 4 years prior. The cutaneous lesions increased in number and size, some showing central necrosis. Her history was otherwise unremarkable. Few cutaneous lesions were present on the head; the palms of her hands, soles of her feet, and mucous membranes were spared.

Findings from laboratory examinations included a high white blood cell count (12.7 \times 10^9/L) with increased neutrophils (8.3 \times 10^9/L), normal values for circulating lymphocytes (2.2 \times 10^9/L) and eosinophils (1.0 \times 10^9/L), and a slight increase erythrocyte sedimentation rate (28 mm/h). Levels for serum immunoglobulin and circulating immunocomplex were normal. Findings from biochemical examination, antinuclear antibody tests, and Waaler Rose test were negative. Findings from hepatitis A, B, and C panels and toxoplasma antibodies were negative. Epstein-Barr virus titers showed positivity only for IgG and Epstein-Barr nuclear antigen, while titers for IgM were negative. Urinalysis showed mild microhematuria and proteinuria. Results of viral, fungal, and bacterial cultures from the vesicles and pustules as well as blood cultures were negative. A skin punch biopsy of 4 mm was obtained from a papular lesion (Figure 3). Antipyretic therapy was administered. Seven days after admission her temperature gradually decreased, and the lesions disappeared after another 7 days with mild residual hypopigmented scars. No recurrences were observed at 1-year follow-up.
Diagnosis and Discussion

Febrile Mucha-Haberman Disease

Figure 1 and Figure 2. Red papules, varicelliform vesicles, and pustules; some evolved into crusted hemorrhagic lesions and others into ulceronecrotic lesions with a sanguineous gray fibrinous exudate and an erythematous halo.

Figure 3. Focal spongiosis and edema accompanied by vascular alteration and neutrophils; necrotic keratinocytes were evident in the epidermis. The cornium layer was orthokeratotic and compact (hematoxylin-eosin, original magnification ×220).

Pityriasis lichenoides (PL) and varioliformis acuta was described in 1916 by Mucha1 and later in 1925 by Haberman2 and is also called Mucha-Haberman disease (M-HD). It is the acute form of PL, an uncommon cutaneous disease mainly occurring in young adult men. Mucha-Haberman disease is not rare in children: 18.6% of the overall population in the series by Romani et al3 were children. In children, M-HD may be only a particular form of PL; in the largest series reported in the literature, most patients had both acute and chronic lesions of PL simultaneously and a chronic course with some acute relapses. The disease M-HD is characterized by the abrupt onset of papulovesicles that evolve into necrotic lesions especially on the trunk (anterior area) and the arms (proximal part and flexor surfaces). Sometimes the eruption involves the entire body surface. Mucous membrane involvement is rare. The lesions are reddish brown papules with central vesiculation and sometimes hemorrhagic necrosis. Usually, the lesions evolve into varioliform scars which leave hypohyperpigmentation.

The eruption is accompanied by only minimal general symptoms. Elevated temperature is a rare complication of M-HD; when fever is present, it is very high (up to 40°C-41°C) and frequently associated with asthenia, severe malaise, and intense myalgias, and the cutaneous eruptions are more severe with painful and large necrotic ulcers. This severe variety first reported by Degos et al4 in 1966 occurs both in adults and children but it appears more frequent in childhood. In children some mucosal lesions have been observed. A central nervous system involvement is described with stupor, agitation, and neuropsychiatric alterations. Arthritis is a common associated complication.

In patients with febrile M-HD, an elevated erythrocyte sedimentation rate, C-reactive protein level, and leukocyte count has been found; in a few cases, antistreptolysin A antibodies and cosinophils were increased. Despite bacteria being isolated from skin cultures, this is not considered the etiological agent of the disease. The histopathological findings of leukocytoclastic vasculitis in the acute variety of M-HD and the occasional presence of circulating immune complexes suggest that the cutaneous lesions are the result of the vascular deposition of an infectious organism, which is the trigger factor of a reaction of hypersensitivity. Other reports suggest an association with a hyperimmune state in M-HD and febrile M-HD.

The course of M-HD is usually self-limited to a few weeks or months, occasionally changing into a chronic course with exacerbations and remissions. The febrile variant may be prolonged for many months and may lead to death. Treatment with antibiotics has been recommended for control of the superinfection. Many other therapies have been proposed; however, no treatment seems completely effective in M-HD. The main treatments proposed in children with M-HD are erythromycin with a 1- to 5-month course, psoralen UV-light treatment or only UV light treatment, and natural sunlight exposition, which all have had good responses in most patients, but the evaluation of their efficacy is difficult because of the tendency of PL to follow a self-limited natural clinical course.

Differential diagnosis between M-HD and other papular or papulovesicular and crusted eruptions of childhood (in particular Crostis syndrome, varicella, and erythema multiforme) may be difficult, and the correct diagnosis is sometimes obtained only by histopathologic analysis. In addition, the transformation into cutaneous T-cell lymphoma observed in sporadic reports of PL5 and the clinical and histopathological similarities between M-HD and lymphomatoid papulosis is still a matter of debate.

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REFERENCES


The Editor is seeking submissions for a new feature, Clinical Problem Solving, which will combine Picture of the Month, Radiologic Case of the Month, and Pathological Case of the Month. Our aim is to demonstrate the thinking process of a master clinician involved in approaching a patient with an unknown disease. The discussion of such cases should place the clinician’s expertise into the context of the prevailing medical literature on the topic. Manuscripts should be between 3000 and 4000 words and may include photographs and radiographs.