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Case Report

A Child with *Neumann* Type Pemphigus Vegetans

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Abstract

Observations: Pemphigus vegetans is very rare in children. There are still no standard recommendations for treatment of childhood pemphigus. Although steroids are often considered to be the first-line treatment, some patients fail to respond. We report a 12 year-old Turkish boy with neumann type pemphigus vegetans. The patient was received 2 mg/kg/day of oral prednisone and azathiopurine 50 mg/day for one month but new lesions still developed. Therefore dapsone 50 mg/day was started and after one week vesicular lesions stopped and skin condition rapidly improved. Azathiopurine was stopped and prednisolone was tapered off. The lesions completely resolved after 4 weeks and dapsone was discontinued after two months. One year after stopping dapsone, the patient showed no new lesions. Cases that do not respond well to steroids, dapsone which is an excellent second-line treatment, may be used for pemphigus vegetans in childhood.

Introduction

Pemphigus vulgaris is an autoimmune disease involving the skin and mucous membranes. Pemphigus is very rare in children [**1, 2**]. Pemphigus vegetans, a variant of pemphigus vulgaris, is the rarest form of pemphigus occuring in %1-2 of all cases [**3**]. To the best of our knowledge, a few cases of pemphigus vegetans have been reported in children [**4, 5**].

Case Report

A 12 year-old Turkish boy presented with a 8-month history of stomatitis. Initially, painful blisters appeared on his tongue and he gradually developed a severe stomatitis. Two months later the lesions had spread to other parts of the body. He complained of an extremely painful mouth and his food intake had diminished. Physical examination revealed thick vegetative, violaceous plaques in the groin, on the axillae, trunk, extremities and perianal region. There were erythematous pustular lesions, some small blisters and erosions on the trunk (**Figure 1**). Ulceration and erosions were present on the oral mucosa. The remainder of physical findings were unremarkable. Investigations showed a white blood cell count of 12540 /µL; eosinophilia and iron deficiency anemia.

Histopathology of a vegetating lesion from extremity showed hyperkeratosis, irregular acanthosis and spongioform abscess formation consisting of neutrophils and eosinophils in the entire epidermis, suprabasal acantholysis, intraepidermal bullae formation. Mixed cell infiltrate was observed in the dermis (**Figure 2**). Direct immunofluorescence (DIF) microscopy showed the deposition of IgG and C3 deposits in intercellular spaces of the epidermis. On the basis of the clinical and laboratory findings, a diagnosis of pemphigus vegetans was made.

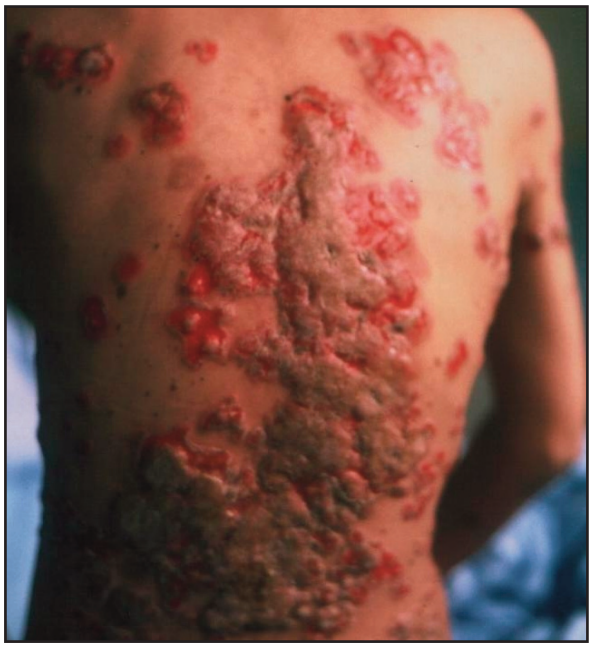

Figure 1. Thick vegetative, violaceous plaques and erythematous pustular lesions, some small blisters and erosions in the groin, on the trunk and extremities.

The patient was received 2 mg/kg/day of oral prednisone and azathiopurine 50 mg/day for one month but new lesions still developed. Therefore dapsone 50 mg/day was started and after one week vesicular lesions stopped and skin condition rapidly improved. Azathiopurine was stopped and prednisolone was tapered off. The lesions completely resolved after 4 weeks and dapsone was discontinued after one month (**Figure 3**). After discontinuation of dapsone therapy for 1 year, the patient showed no new lesions.

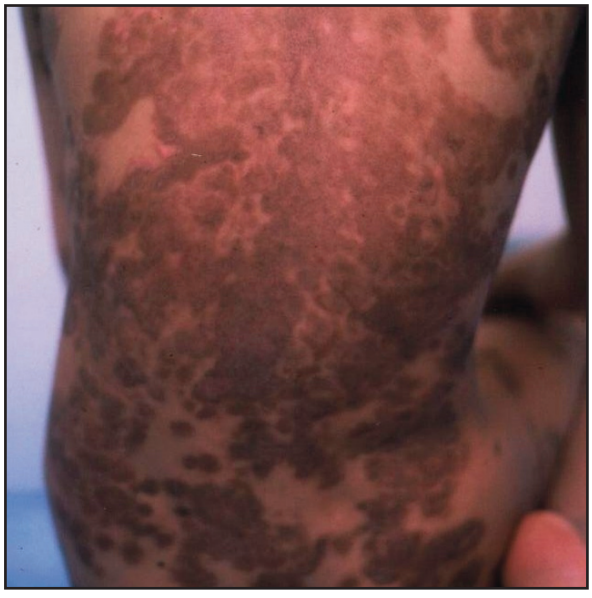

Figure 3. The lesions healed slowly over the next four weeks and left hyperpigmented macules after treated with dapsone

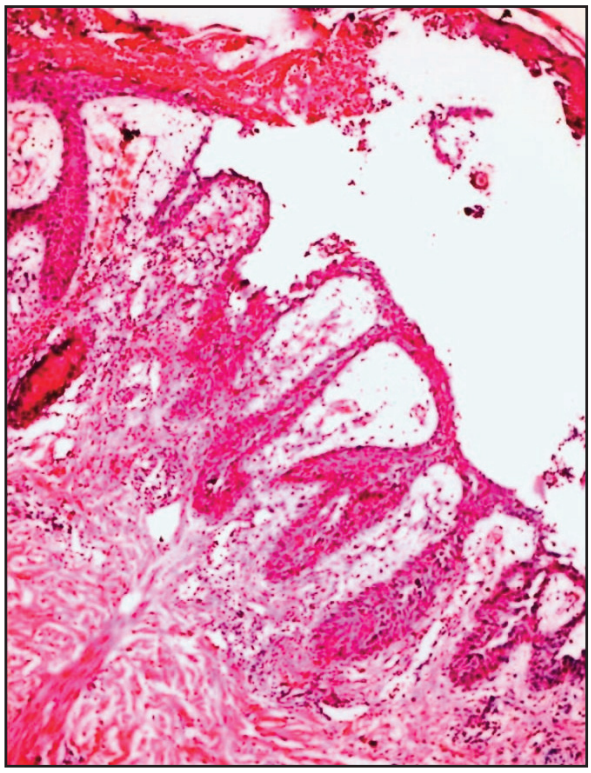

Figure 2. Lesional biopsy specimen shows hyperkeratosis, irregular acanthosis and spongioform abscess formation consisting of neutrophils and eosinophils in the entire epidermis; suprabasal acantholysis, intraepidermal bullae formation and mixed cell infiltrate in the dermis. HE x 100

Discussion

Pemphigus vegetans is considered to be a rare variant of pemphigus and characterised by blisters and erosions associated with verrucous vegetations [**6**]. Pemphigus vegetans has two clinical subtypes; the *Neumann* type described in 1886, and the *Hallopeau* type, described in 1889. The *Hallopeou* type starts with circumscribed pustules, which histologically shows eosinophilic intraepidermal pustules. It has a relatively benign course, require lower doses of systemic corticosteroids and usually have a prolonged remission. The *Neumann* type is seen more commonly but develops extensive lesions that are refractory to medical treatment. It needs higher doses of systemic corticosteroids, and has relapses

and remissions. The lesions in the *Neumann* type begin as vesicles and erosions. The mean age of onset in *Neumann* type is 44 years and in *Hallopeau* type is 45 years according to the analysis of the reported cases [**7**].

The most important differential diagnosis is the pyodermatitis-pyostomatitis vegetans which show similar clinical and histological findings, however, the immunflorescence results characteristically negative. Halogenederma, blastomycosis-like pyoderma, pyoderma gangrenosum also must be excluded [**8**].

Although there are several reports of pemphigus vulgaris and pemphigus foliaceus in childhood, we have found only a few cases with juvenile onset pemphigus vegetans [**4, 5**]. The first case has been reported previously, that of a 12-year old Chinese boy treated with traditional Chinese herbal medicine [**5**]. The second one was a 14-year old Thai boy treated with oral prednisolon in combination with azathiopurine [**4**]. The third one was a 7-year old boy who developed pemphigus vegetans shortly after a second liver transplantation. His condition had developed while being treated with immunosupressive drugs that are used to treat immunobullous disease. DIF examination of skin revealed prominent IgA intercelular staining of the epidermis in a pemphigus-like patern and he was treated with dapsone [**9**].

The prognosis in childhood pemphigus is considerably variable [**4**]. Systemic corticosteroids are the mainstay of therapy [**10, 11**]. Oral prednisolone alone or in combination with adjuvant therapy, which is used for steroid-sparing effect, has been used with good results. Immunosuppresives such as azathiopurine, methotrexate and cyclosporine, gold, antimalarials and dapsone are choises for adjuvant therapy [**4**]. When it comes to a persistent verrucous vegetation form, the combination of corticosteroid and etretinate resulted in healing of the lesions [**11**].

This case is found worth to be presented for both displaying a rare form of pemphigus and for the relatively low incidence of the disease in childhood. Dapsone has achieved excellent results in our case but there are still no standard recommendations for treatment of childhood pemphigus.

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