

Complete recovery from juvenile pemphigus vulgaris

Sir,

Pemphigus is an uncommon mucocutaneous disease caused by autoantibodies against desmosomal antigens. Juvenile cases are rare. The diagnosis is often delayed due to confusion with other entities.

An 11-year-old girl without any previous medical problems

had one month history of flaccid bullous lesions and large crusted hyperkeratotic erosions on the trunk, limbs, and face without mucosal localization. There was no family history of autoimmune processes. No improvement was noted with a well-conducted antibiotic treatment.

A lesional biopsy showed suprabasal acantholysis and direct immunofluorescence (DIF) studies of frozen skin tissue showed positive intercellular staining for IgG within the epidermis. Blood samples for indirect immunofluorescence (IIF) on rabbit's lips demonstrated circulating IgG autoantibodies at a titer of 1:100.

A diagnosis of juvenile pemphigus vulgaris (PV) was made. Treatment was started with 1 mg/kg/d (50 mg/d) methylprednisolone. A gradual remission was observed. Complete regression of lesion was obtained after 30 days, and the dose of methylprednisolone was gradually reduced after 45 days without any recurrence. No side effect was observed.

After 3 years the maintenance therapy was discontinued, and the girl has had no relapses over the last 7 years. Unfortunately, this remission could not be confirmed by DIF and IIF.

Pemphigus is a group of autoimmune blistering skin disease characterized by blister formation. Blisters are due to loss of keratinocyte cell–cell adhesion in the superficial and deep epidermis respectively.^[1] The incidence rate of pemphigus in Tunisia is 6.7 cases per million per year. High rates of pemphigus foliaceus (PF) among young people living in rural areas are reminiscent of Brazilian pemphigus. However, the absence of cases among genetically related household members and during childhood, as well as the large predominance among women, contrasts with Brazilian pemphigus.^[2]

Juvenile pemphigus, except the endemic form, is rare.^[3] Only 69 cases of PV and 19 cases of PF have been reported. Stomatitis is the presenting sign in more than 50% of the children with PV. In our case, lack of mucosal changes may suggest cutaneous pemphigus foliaceus. But in some children, skin blisters may be the single symptom of PV, and no mucous membrane lesions are present.^[4] Early diagnoses in our patient (after 1 month) have probably prevented mucosal localization.

In children, pemphigus may be misdiagnosed as bullous impetigo; other blistering diseases more common in

children are dermatitis herpetiformis, IgA linear dermatosis, epidermolysis bullosa, and Stevens-Johnson syndrome. It requires a high index of suspicion in order to make an early diagnosis and to avoid treatment delay.

Systemic corticosteroids are the treatment of choice for pemphigus vulgaris. Immunosuppressive agent (azathioprine, cyclophosphamide) could be added for patients with severe disease that cannot be controlled by corticosteroids alone or to reduce the dose of corticosteroids.^[5] However, immunosuppressants continue to have the complications of systemic infections. Successful use of rituximab therapy has been reported in refractory childhood pemphigus vulgaris.^[6] Some authors recommend use of intravenous immunoglobulins for cases of childhood and juvenile pemphigus, in which this therapy can delay the need for administration of immunosuppressive drugs.^[7]

Prognosis of pemphigus is usually better in childhood than in adulthood, except for paraneoplastic pemphigus (with little data on children); but PV also seems to show a relapsing course in the pediatric age group like in adults. However, as seen in our case, complete recovery is also possible. Our case is only the second such completely recovered case reported in the literature.^[8]

N. El Fekih, M. Kharfi, A. Karaa, M. R. Kamoun

Department of Dermatology, Charles Nicolles Hospital, Tunis, Tunisia

Address for correspondence: Dr. N. El Fekih, Department of Dermatology, Charles Nicolles Hospital, Boulevard 9 avril Bab Saadoun Tunis – Tunisia. E-mail: fekih.nadia@planet.tn

REFERENCES

1. Mahoney MG, Wang Z, Rothenberger K, Koch PJ, Amagai M, Stanley JR. Explanations for the clinical and microscopic localization of lesions in pemphigus foliaceus and vulgaris. *J Clin Invest* 1999;103:461-8.
2. Bastuji-Garin S, Souissi R, Blum L, Turki H, Nouira R, Jomaa B, *et al* Comparative epidemiology of pemphigus in Tunisia and France: Unusual incidence of pemphigus foliaceus in young Tunisian women. *J Invest Dermatol* 1995;104:302-5.
3. Jun H, Antaya RJ. Pemphigus vulgaris in an adolescent. *Curr Opin Pediatr* 2002;14:426-8.
4. Yoshida K, Takae Y, Saito H, Oka H, Tanikawa A, Amagai M, *et al*. Cutaneous type pemphigus vulgaris: A rare clinical phenotype of pemphigus. *J Am Acad Dermatol* 2005;52:839-45.
5. Gupta MT, Jerajani HR. Control of childhood pemphigus erythematosus with steroids and azathioprine *Br J Dermatol* 2004;150:163-4.
6. Kong HH, Prose NS, Ware RE, Successful treatment of refractory childhood pemphigus vulgaris with anti-CD20 monoclonal antibody (rituximab). *Pediatr Dermatol* 2005;22:461-4.
7. Szép Z, Danilla T, Buchvald D. Treatment of juvenile pemphigus vulgaris with intravenous immunoglobulins. *Cas Lek Cesk* 2005;144:700-3.
8. Harangi F, Varszegi D, Schneider I, Zombai E. Complete recovery from juvenile pemphigus vulgaris. *Pediatr Dermatol* 2001;18:51-3.