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Pediatric Pemphigus

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Pediatric Pemphigus

Pemphigus is a group of autoimmune blistering disorders characterized by blister formation that is caused by loss of keratinocyte cell-to-cell adhesion in the epidermis due to circulating autoantibodies against desmosomal proteins [1]. The disease is extremely rare in pediatric age group. Pemphigus vulgaris (PV) is the most common form seen in children, as seen in adults. Although prevalence of pemphigus in this age group is unknown, childhood PV accounts for approximately 1.4-3.7% of total cases [2,3].

Pemphigus, a life-threatening autoimmune vesiculobullous disease, is characterized by Nikolsky's positive flaccid blisters, painful and persistent erosions and crusts in the skin [1,4]. The mucous membrane is the first affected site in majority of patients. Oral mucosa is the most frequently affected mucosal site, with painful and readily bleeding erosions and ulcers. Other mucosal membranes such as genital, ocular, nasal mucosa are less frequently involved. The clinical manifestation of pemphigus is not similar in children and adult patients. Interestingly, frequency of involvement of the genital and ocular mucosa is reported to be significantly higher in children than adult PV. Epistaxis and hoarseness can be seen owing to involvement of the nose, pharynx, and larynx. Due to pain, food and fluid intake may be disturbed, child may complain of difficulties with urination or defecation [5].

The diagnosis is often delayed due to rarity of pemphigus in this age group and confusion with other entities. So, high index of suspicion is required in order to make an early diagnosis. Differential diagnosis of cutaneous lesions include bullous impetigo, herpetic infections, staphylococcal scalded skin syndrome, bullous contact dermatitis, bullous erythema multiform, Stevens–Johnson syndrome, inherited forms of porphyria, and congenital blistering disorders (epidermolysis bullosa) and other autoimmune bullous diseases (chronic bullous disease of childhood, bullous pemphigoid, dermatitis herpetiformis, IgA linear dermatosis). Sexual abuse, bullous lichen sclerosis and bullous fixed drug eruption should be considered in the differential diagnoses of genital lesions and acute herpetic gingivostomatitis, hand-foot-mouth disease, oral candidiasis, erythema multiform, other autoimmune bullous diseases (linear IgA disease, cicatricle pemphigoid, bullous pemphigoid of childhood and paraneoplastic pemphigus), Behcet's disease, aphthous stomatitis, lichen planus should be considered in the differential diagnosis of oral lesions [1]. Histopathological and immunofluorescence examinations are very important in the diagnosis of pemphigus. Acantholysis and intraepidermal blister formation are characteristic findings on histopathological examination. Direct immunofluorescence shows the deposition of IgG around keratinocytes in a "chicken wire" or "crazy paving" pattern. The detection of circulating anti-epidermal antibodies in the serum of patients with PV by indirect immunofluorescence further supports the diagnosis. The antibody titers correlate with activity of disease in some patients and can be used for follow-up [1].

Since pemphigus is rare in pediatric population, evidence-based treatment guidelines have not been reported yet. Immunosuppression is the mainstay of therapy and systemic corticosteroids are the treatment of choice (prednisolone 1-2 mg/kg/day) to control the disease during the acute phase [1]. In pediatric age group, the dose should be adjusted according to age, body weight, the severity of the condition and the side-effects of the drug. When the disease begins to go into remission, the dose can be tapered slowly. The continuation of treatment for long time is not preferred due to long-term side-

effects of systemic steroids. Other immunosuppressive agents (azathioprine, cyclophosphamide, methotrexate, cyclosporine, and micophenolate mofetil) and steroid-sparing agents (dapsone, sulfa pyridine, gold, and erythromycin, antimalarial) can be used concomitantly for allowing a decrease in the dose of corticosteroids required to control the disease. Among these drugs, dapsone and azathioprine are the most commonly used adjuvants for pemphigus treatment [3]. Potent topical or intralesional corticosteroids can be used for localized disease or isolated recalcitrant foci [1]. Intravenous immunoglobulin (IVIG) is a safe and effective treatment option in pemphigus and is recommended by some authors in pediatric age group due to decrease the need for administration of immunosuppressive drugs [3].

Rituximab, a monoclonal antibody directed against the CD20 antigen on B lymphocytes, is promising in refractory cases. This drug is usually used in conjunction with immunosuppressant or high-dose IVIG. In children, rituximab-related decrease in immunoglobulin levels is one of the frequently seen side effects, so in order to reconstitute normal immunity combination therapy with IVIG is recommended in pediatric cases [4]. Plasmapheresis and immunoadsorbption can be effective in severe cases [3].

Pemphigus generally shows a relapsing course in the pediatric age group like in adults and complete recovery is rare. Children with pemphigus show a relatively better prognosis when compared with adults, except for Para neoplastic pemphigus [1]. High-dose and prolonged administration of CS and other adjuvant steroid sparing agents may result in serious side effects (such as infections, Cushing disease, hypertension, cataract, growth retardation, osteopenia, diabetes mellitus, avascular necrosis, etc.) so an important part of pemphigus morbidity is related with the treatment [4]. In a retrospective analysis of 33 childhood PV cases, it was reported that serious side effects developed in 60.6% of patient (cushingoid features (65.0%), growth retardation (50.0%), and infection (50.0%)) and one patient died due to infection [5]. Majority of adult PV patients requires longer use of these therapies than children for controlling disease, so more severe immunosuppression occurs in adults. Due to this prolonged immunosuppression and comorbidities, the mortality rate in adults is higher than in children (10-15% vs 2.9%, respectively) [5].

Conclusion

The diagnosis of pemphigus in children can be delayed due to the rarity of pemphigus in this age group, so high index of suspicion is necessary for accurate diagnosis. The prognosis of disease seems to be better in childhood when compared with adults. The treatment of pemphigus is similar in children and adults but clinicians should remember that children during periods of growth and development are extremely vulnerable to the side effects of systemic corticosteroid treatment.

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