JUVENILE PEMPHIGOID OR DERMATITIS HERPETIFORMIS

A Case Report

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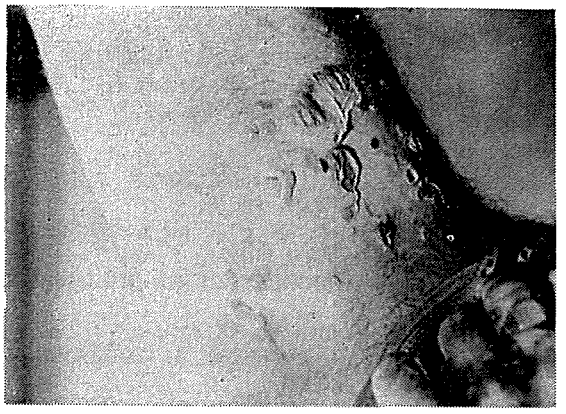
The condition is an uncommon widespread bullous eruption occurring in children, with a predilection for the orifices of the face and for the genitocrural region. It differs in some respects from both the adult variety of dermatitis herpetiformis and pemphigoid.

Case Report:

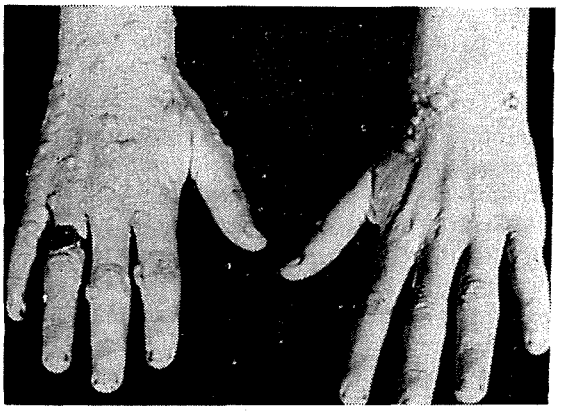
The patient was a twelve year old boy, presenting with an extensive blotchy rash initially accompanied by sparse bullae. He had arrived from Australia two weeks before, where he had received vaccination one week before departure. He also had dental extraction under local anaesthesia a week before the rash occurred. There was no history of any drugs or of any previous skin complaints. No irritation or itching or any constitutional disturbances accompanied the rash. On admission to hospital the bullae continued to increase in size and number within the erythematous patches over the next few days, but the patient's general condition remained good..

On examination the initial rash consisted of polycyclic, gyrate, erythematous plaques, covering almost the whole body, including the face, palm and soles, and containing tense vesicles and bullae. The bullae increased in size and number, and were either yellow, containing clear serum, or haemorrhagic in nature. Nikolsky's sign (or shearing of epidermis on slight sliding pressure of fingers occurring when bullae are intraepidermal) was



negative, confirming together with the tense consistency, that the bullae were subepidermal. The bullae were concentrated on the neck and face, where vesicles presented around eyelids, nasal vestibules and lips, genitocrural areas (Fig. 1). buttocks, dorsum of hands and feet (Fig. 2), and extensor aspect of knees and elbows. The buccal mucosa and palate showed also some vesicles. The new bullae and vesicles arose at the periphery of the gyrating plaques, forming the characteristic 'cluster of jewels' appearance, and some bullae, especially in the genitocrural areas, coalesced to form an arcuate pattern. The eruption resembled bullous erythema multiforme, but there were no target lesions and no clinical evidence of any skin necrosis. The cluster of vesicles around the facial orifices and of bullae on the genitocrural regions favoured the diagnosis of juvenile pemphigoid.

Biopsy of a vesicle from the back showed separation of the epidermis from the dermis with fibrin accumulation. The dermis showed a sparse leucocytic infiltrate, but no eosinophils, with conspicuous intercellular oedema. There were no papillary microabscesses characteristic of adult dermatitis herpetiformis, and the epidermis showed no necrosis as in erythema multiforme. Other investigations, including a full blood count and picture, blood urea, urinalysis, and liver function tests, were normal.



The patient was treated with emollient baths with emulsifying ointment, dilute potassium permanganate compresses applied frequently to the bullae, and the application of chlortetracyline ointment to the vesicles around the facial orifices to prevent secondary infection. Dapsone was started orally in a daily dosage of 150mg. There was a marked improvement within 48 hours, the bullae decreasing in size with no new formation, and the rash subsiding gradually within a week. After a fortnight, the dapsone was reduced to 100mg. daily for another 3 weeks, then 50 mg. daily for another 3 weeks, stopping it after another 4 week period of 50mg. alternate days. There was no recurrence after stopping the drug, the patient being followed up for a year. Patient had a weekly full blood count and picture to detect any haemolytic anaemia or agranulocytosis resulting from the dapsone. Once the serum from the bullae was reabsorbed, their thick roof rested flaccidly on the raw bases, acting as a natural graft for rapid healing without any scar resulting. Some milia (or inclusion epidermal cysts) common after resolution of subepidermal bullae, were present over knuckles, knees and elbows.

Discussion:

Dermatitis Herpetiformis (D.H.) is not an uncommon disease, occuring in middle age, but it is rare in children. It has been said that two forms exist in children, a bullous variety and the classical papular or papulovesicular type. The former is the classical juvenile pemphigoid.

Early accounts of D.H. in children were by Bowen (1901, 1905) and Knowles (1907). Bowen described 15 cases, six of which followed vaccination similar to the case described by Morris (1896). However, usually there are no apparent precipitating factors. Knowles drew attention to the essential differences between the form in children and that in adults. Itching, the typical morphology and patchy distribution, and the postinflammatory hyperpigmentation, so constant in the adult variety, are usually absent. Points of similarity are the periods of remission and relapse, the presence occasionally of blood eosinophilia, and the dramatic response to dapsone or sulphayridine (a true therapeutic test.) Knowles found that relapses decreased with age and finally completely cleared by puberty, so that the prognosis is better than the adult type. Peterkin (1951) reviewed 105 cases of D.H. seen in Edinburgh, and these included 21 childhood cases of the bullous type. In the latter, the prognosis was good, the case clearing by puberty, wheras in adults the disease was chronic.

The condition is not accepted by all as D.H. Tolman et al (1959) expressed the view that it is probably a form of erythema multiforme. Similar views were expressed by Katz (1968) and Lever (1957), the reasons given for this opinion being the lack of symmetrical distribution and grouping of the rash, the predominance of bullae rather than vesicles or excoriated papules, frequent mucous membrane involvement, absence of pruritus, or any postinflammatory pigmentation, and an inconsistent response to dapsone or sulphapyridine. However, Grant (1968) states that the mucous membrane lesions are rare, there are no definite apparent predisposing factors as in erythema multiforme, and the response to treatment and the histological picture are different. Ingram (1960) holds that it is D.H., the variation in the clinical picture being merely a question of difference in skin reaction before puberty. Smith (1966) and Sneddon (1968) describe the bullous variety under the title of juvenile pemphigoid, and state that it is different from D.H. Grant (1968) reviewed 38 cases, and agrees that the condition is akin to pemphigoid but not true pemphigoid. From this brief review of the literature, it is seen that there is no general agreement as to the true nature of this characteristic prepubertal bullous eruption.

Juvenile D.H. or pemphigoid has an acute onset without any constitutional disturbances, or if present, consisting of mild pyrexia, malaise, and anorexia. The eruption consists of tense subepidermal bullae, 1 or 2 cm. in diameter, arising on polycyclic, erythematous, urticarial plaques. Characteristically, the bullae arise in crops, usually confined to the periphery of the gyrating plaques, and compared to a 'cluster of jewels pattern'. There is no herpetiform grouping, the bullae coalescing to form an annular or arcuate pattern. Some bullae are haemorrhagic instead of having a clear serum content. Once ruptured or after spontaneous reabsorption, the bullae heal rapidly and rarely become impetiginised or eczematised. There is a minimum tendency for pigmentation or scarring, some milia remaining over the extensor aspect of some joints. The eruption is asymptomatic.

The distribution in descending order of frequency (Grant (1968), includes the genitocrural regions, buttocks, thighs, lower abdomen commonly, then the face and neck, hands and feet, and around joints, the least commonly affected being the mucous membranes. The bullae arise in crops and a generalised eruption is rare. The distribution differs from that of adult D.H. Onset starts in 50% of cases before the age of 5, usually between 3 and 4 years. Boys are more commonly affected. The disease remits with therapy within a few weeks and relapses can occur over an average period of 3 years. Cases of relatively late onset, between 9 and 12 years, have a better prognosis, in that relapse, after adequate therapy for a three month period, is rare.

Histology shows the bullae to be subepidermal, lymphocytes polymorphs with sparse eosinophils, being present in the fibrinous material of the blister fluid. Epidermis is intact with no necrosis. The

picture resembles pemphigoid seen in elderly patients, as the pathognomic microabscesses in the dermal papillae of adult D.H. are rarely seen.

Blood eosinophillia is common as in adult D.H., but all other investigations are normal. Differential diagnosis in young children includes impetigo, epidermolysis bullosa, incontinentia pigmenti, and acrodermatitis enteropathica. In older patients, erythema multiforme, papular urticaria, urticaria pigmentosa and drug eruptions have to be considered. Diagnosis is made on the clinical picture and the dramatic response to specific treatment.

Dapsone and sulphapyridine constitute specific therapy, especially the latter (Grant 1968) if a large initial dose is given. Sulphapyridine is less toxic than dapsone in children. Spontaneous remissions have also been recorded. The prognosis is better than the commoner adult bullous diseases.

Summary:

A case of juvenile pemphigoid or dermatitis heretiformis is described. Oral lesions were present. There was a therapeutic response to dapsone therapy for a three month period, with no relapse after a one year follow up. Vaccination may have been the predisposing factor.

References

BOWEN, J.T. (1901, 1905). J. Cutan. Dis. 19, 401; & 23, 381.

GRANT, P.W. (1968). TRANS. of the St. John's Hosp. Derm. Soc. 54, 128.

INGRAM, J. T. (1960). Brit. J. Derm. 72, 60.

KATZ, H. (1968). Arch. Derm. 97, 90.

KNOWLES, F. C. (1907). J. Cutan. Dis. 25, 247.

LEVER, W. F. (1957). Acta Dermat. Vener.

MORRIS, M. (1896). Brit. J. Derm. 8, 278.

MORRIS. M. (1896). Brit. J. Derm. 8, 278.

PETERKIN, G.A. (1951). ibid. 63, 1.

SMITH, E.L. (1966). Trans. St. John's Hosp. Derm. Soc. 52, 176

SNEDDON, I.B. (1968). Textbook of Dermatology (Rook, A.J., Wilkinson, D.S. & Ebling, F. J. G.). 2, 1187.

TOLMAN, M.M., MOSCHELLA, S.L., SCHNEIDERMAN , R.N. (1959). J. Invest. Derm. 32, 557.