

The hair itself was carefully examined. It was white, sparse, short (less than 1 centimetre), very brittle, and looked like down. On optical light microscopic examination, the hair did not show any evident anomaly, but only slight, non-specific changes. On the other hand, we were surprised to find very characteristic anomalies with the polarizing microscope; indeed, examination with polarized light revealed a very specific defect, not reported previously, including the following numerous and simultaneous aberrations: (1) the ends of the hair are spindly, moniliformis, sharp at the distal end, "spearhead-like"; (2) at the proximal end of the spindle, there is a narrow and deficient area which gives rise to an elegant wave; (3) sometimes there is another elegant wave, lying in the opposite direction, thus giving a general swan-necked appearance; (4) furthermore, the whole shaft displays numerous striae, disposed in little groups of three or four, which correspond to areas of brittleness—a kind of mild trichonodosis, or slight trichoschisis.

The juxtaposition of these very strange aberrations probably renders the patient's hair a specific hair defect, an indicator of the disease, directly related to zinc deficiency. Such findings have not been reported previously.

Successful results were obtained with zinc therapy. Within a month the eruption resolved, the diarrhea stopped and the growth of hair was gradually restored. Three months later a new examination with polarized light showed a quite normal structure of the hair shafts: all the previously observed anomalies had disappeared.

Chronic Bullous Dermatitis of Childhood

C. Del Forno, A. Giannetti and G. Orecchia

*Department of Dermatology, University of Pavia,
Pavia, Italy*

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Abstract. A case of chronic bullous dermatitis of childhood in a 3-year-old boy is described. Immunofluorescence tests were negative and biopsy of the jejunal mucosa showed marked villous atrophy. The dermatosis

was brought under control by a combination of diamino-diphenylsulphone and systemic steroids. The relationship with other bullous eruptions of childhood such as dermatitis herpetiformis and bullous pemphigoid is discussed.

The chronic acquired bullous dermatoses of childhood include: pemphigus vulgaris, bullous pemphigoid (BP) and dermatitis herpetiformis (DH), with their typical clinical, histological and immunological features. Moreover, various cases of bullous dermatosis have been described, having a clinical picture almost exclusively of large blisters, with a histological picture of subepidermal bullae and generally with negative immunofluorescence (IF) tests, responding variably to diaminodiphenylsulphone (DDS) or sulphapyridine. For this disease the name chronic bullous dermatosis of childhood (CBDC) was proposed (7).

However, some authors prefer to use the term juvenile DH (5, 2) even in the absence of the typical immunological findings. On the other hand, review of the literature suggests that some cases described as CBDC should be classified as DH, because they have IF features typical of DH: deposits of IgA at the dermo-epidermal junction or at the tip of the dermal papillae (1, 8).

As far as intestinal abnormalities in blistering diseases of childhood are concerned, the atrophy of the jejunal mucosa, which is present in most cases of DH (3), was found in cases described as BP of childhood (9), or juvenile DH (10); in these cases IF tests were not carried out. Other authors found a non-specific inflammatory reaction in the jejunal mucosa (5), or abnormalities of gastrointestinal functions (2) in some patients with a clinical picture of juvenile DH.

We now report a case of CBDC in which it was possible to perform the biopsy of the jejunal mucosa and the IF tests.

CASE REPORT

A 3-year-old boy was admitted to our Department in July 1977 with an extremely pruritic widespread bullous dermatosis, which started in January 1977. The initial lesions—vesicles and blisters with secondary erosions and crusts—appeared on the pinna. Subsequently lesions appeared on the lower extremities, buttocks, penis and abdomen. The dermatosis ran a chronic course. In May 1977, after admission to a local hospital, a diagnosis of juvenile pemphigoid was made on the basis of clinical data and a Tzanck smear test. He was treated with sulpha-



Fig. 1. Numerous large bullae and crusted lesions on the back.

methoxypyridazine 500 mg daily per os, with partial improvement, followed by a relapse after a few weeks.

At the time of admission to our Department the child was well developed and in good general health, but distressed by the severe pruritus. The past medical history was normal except for an episode of diarrhea at 5 months and febrile convulsions at 12 months. On physical examination, multiple bullous lesions were evident on the head, lower trunk, external genitalia and, to a lesser extent, on the extremities (Fig. 1). The mucous membranes were spared. Most of the lesions were large, tense blisters, filled with clear fluid; there were also some smaller vesicles grouped in a herpetiform manner, on normal, sometimes slightly erythematous, skin.

Histological examination of a blister biopsy sample showed a subepidermal bulla, with accumulation of

neutrophils in the papillary dermis and a moderate inflammatory infiltrate in the subpapillary dermis (Fig. 2). Direct IF performed on three biopsies taken at different times on new lesions and adjacent skin failed to show any deposits, either of immunoglobulins (IgG, IgA, IgM) or of complement; circulating autoantibodies were not found by indirect IF. Iodine patch test performed with both tincture of iodine (Pharmacopaea Italica) and potassium iodide 25% in petrolatum were negative.

Laboratory data showed the following abnormalities: transient low values of serum iron, eosinophilia (up to 1800 eosinophils/mm³) and abnormal xylose and lactose tolerance tests. Intestinal biopsy, performed at the Paediatric Department "G. & D. De Marchi" of the University of Milan, Italy, showed marked villous atrophy of the jejunal mucosa.

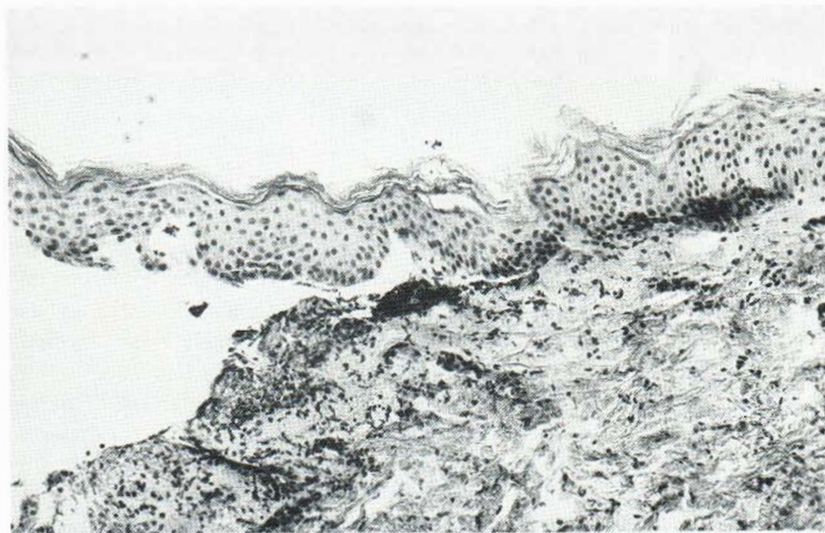


Fig. 2. Subepidermal bulla with moderate inflammatory infiltrate in the upper dermis ($\times 160$).

Treatment was started with DDS, 50 mg per os daily. It was well tolerated but led to only partial control of the lesions and pruritus. Complementary systemic steroid therapy with betamethasone, 1.5 mg per os daily, led to complete disappearance of the symptoms within few days. Good control was maintained after reduction of the dose of betamethasone to 0.5 mg on alternate days, but attempts to discontinue the steroid were unsuccessful because of the rapid recurrence of blisters and pruritus. Since January 1978 the child has been on a gluten-free diet (which according to his mother he observes strictly). He is presently receiving 50 mg of DDS daily and 0.5 mg of betamethasone every second day. Occasionally 2-4 bullae have appeared, usually when the betamethasone dose is reduced to 0.25 mg every other day.

DISCUSSION

On histological examination of this case of blistering disease a subepidermal bulla was seen, as characteristic of BP and DH, but the negative IF tests performed on skin and serum led to exclusion of these potential diagnoses and a diagnosis of CBDC was therefore considered, although jejunal biopsy showed villous atrophy as in DH and functional tests showed the presence of intestinal malabsorption. Now some authors (6) believe that after a certain length of time most cases of CBDC evolve into true cases of DH, with the typical IF findings; in the present case the finding of villous atrophy of the present case the finding of villous atrophy of the able. However, because previously positive IF tests were observed in all the 22 cases of DH (15 children and 7 adults) seen in our Department (4), because DDS alone failed to control the disease and because after 7 months of gluten-free diet it was impossible to reduce the dosage of the drugs, this case was classified as CBDC.

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Multiple Glomus Tumours: A Report of a Family in Denmark

Finn Schultz Larsen and Esther Hage

Departments of Dermatology and Pathology, Odense University Hospital, Odense, Denmark

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Abstract. In a family affected by multiple glomus tumours, one of the members had approximately 500 tumours spread over the entire skin. The histopathological similarity to cavernous haemangioma is emphasized. We consider that multiple glomus tumour is a more common skin disease than is assumed today.

Multiple glomus tumour (MGT) is a benign, dominant, hereditary skin disease. The tumours develop from specialized arterio-venous anastomoses, the glomus bodies, which are particularly numerous on the distal parts of the extremities. Only 21 families suffering from MGT (3, 4, 5) have been reported since Touraine (6) first described the disease in 1936. The clinical picture is characterized by painless, small bluish, partly compressible tumours, often somewhere between 10 and 20 in number, on the extremities. We have studied a family affected by MGT, where one of the members had approxi-