Treatment: L-thyroxine 0·1 mg daily caused the loss of 4 kg during the first month and some thinning of the ankles. The application of betamethasone valerate ointment removed the scaling but did not influence the thickening. Injection of triamcinolone acetonide suspension on three occasions had no effect.

Investigations: Biopsy of skin of leg showed the typical appearance of circumscribed myxœdema, with large amounts of mucoid material staining well with Alcian blue, which extended throughout the dermis, widely separating the collagen bundles. The epidermis showed marked hyperkeratosis with follicular plugging.

Radio-iodine studies before operation showed hyperthyroidism, after operation borderline hypothyroidism.

X-rays show cortical thickening of both first metacarpals and metatarsals. The new bone is laminated and particularly on the right first metacarpal has a characteristic 'soap-bubble' appearance. The terminal phalanges, radii and ulnæ are unaffected; this fact and the nature of the new bone are features distinguishing the changes from pulmonary hypertrophic osteoarthropathy. The findings are typical of thyroid acropachy.

Serum cholesterol, serum calcium, hæmoglobin estimations normal. Serum protein bound iodine 3·4 µg/100 ml.

Comment

This case is presented as a typical example of a rare syndrome. Malkinson (1963) found only 26 authentic cases in the literature, including the 6 of Gimlette (1960) from St Thomas's Hospital. No case seems to have been presented in this Section, but Peard (1961), Summerly (1962) and Sweet (1962) have shown cases at the Endocrinology Section of this Society, and at the North of England and at the West of England and Wales Dermatological Societies respectively.

REFERENCES
Gimlette T M D (1960) *Lancet* i, 22
Malkinson F D (1963) *Arch. Derm., Chicago* **88**, 303
Peard M C (1961) *Proc. R. Soc. Med.* **54**, 342
Summerly R (1962) *Brit. J. Derm.* **74**, 105
Sweet R D (1962) *Brit. J. Derm.* **74**, 381

The following cases were also shown:

Ulcerative Colitis with Bullous Eruption
Dr Harold Wilson
(1) Dermatitis Gangrenosum Infantum
(2) Epidermolysis Bullosa Dystrophica (Hurlitz)
Dr E J Moynahan
Lupus Erythematosus
Dr E L Rhodes (for Dr R H Marten)
Case for Diagnosis – Telangiectatic Erythema of the Face
Dr R H Marten
Dermatomyositis
Dr J Warner (for Dr D I Williams)
Multiple Cutaneous Nodules
Dr J Warner (for Dr R H Marten)
Mycosis Fungoides with Dermatitis Herpetiformis or Pemphigoid
Dr M L Johnson (for Dr P J Hare)
Mycosis Fungoides
Dr K V Sanderson
Kaposi's Hæmorrhagic Sarcoma treated with Intra-arterial Mustine
Dr F Bor (for Dr M Feiwel)
Infiltrated Plaques of the Scalp
Dr A Bowyer (for Dr Louis Forman)
Multiple Pyogenic Granuloma
Dr I Sarkany

Meeting January 21 1965

Generalized Pustular Psoriasis (von Zumbusch) with Episodic Hypocalcæmia

P W M Copeman MRCP and A M Bold BM
(for H J Wallace FRCP)

R G, male, aged 51. Retired artesian well borer

History: He presented five years ago with generalized guttate psoriasis two weeks after tonsillitis, coryza, and bilateral orchitis (? mumps). After three months the eruption became pustular and he was admitted with rigors, prostration, leucocytosis and ESR 30–50 mm in 1 h (Westergren).

Family history: Not relevant.

Past history: No operations. 1951, contact dermatitis (oil) hands and legs.

Course: He was admitted on four subsequent occasions, for two to four months, at intervals of two to fifteen months, with acute relapse of generalized pustular psoriasis; the fever and severe systemic disturbance reflected the condition of the skin.

Each exacerbation had similar features: (1) Infection: The severe attacks appear to be heralded by an infection, usually of the upper respiratory tract. It was associated with orchitis (1960) and chronic prostatitis (1961); there has been no urethritis. No drugs have been implicated. (2) Fever and rigors: Temperature 100–103° F, lasting for one to four weeks. Very severe systemic disturbance; depressed, stuporous. (3) Arthralgia: Occasional swelling of large joints. (4) Generalized eruption: A rapidly spreading erythema with

pustulation and exfoliation, not itchy, affecting scalp and usually face, trunk and limbs, particularly severe in groins; finger and toe nails shed; occasional stomatitis and conjunctivitis. The eruption was either generalized with small, often coalescent, sheets of pustules leaving few unaffected areas of skin, or polycyclic and erythematous with central clearing, the lesions being surmounted by small pustules.

Between attacks the patient is fit and the skin is almost clear, although more recently the pustular psoriasis of palms and soles has been particularly troublesome.

Treatment: Local: a variety of antibiotic and corticosteroid preparations. Systemic: several antibiotics, vitamins, folic acid, and dapsone. Calcium lactate 2 g and calciferol 50,000 units daily since December 1962. He has not had antimetabolites or systemic corticosteroids. Treatment did not appear to influence the course of the disease.

Investigations: Histology from several lesions of different morphology (Dr I Whimster and Dr G C Wells): 'Pustular psoriasis – subcorneal and intraepidermal microabscesses'. Precipitating infection (viruses, mycoplasmata) not identified (Dr A Dudgeon and Dr C S Nicol). Unbroken pustules sterile.

Biochemistry: The main biochemical abnormalities were: low serum calcium and plasma protein concentrations; a high plasma alkaline phosphatase and increased fæcal fat excretion. In each of five admissions to hospital with generalized pustular psoriasis, the serum calcium was initially low (6·9–8·2 mg/100 ml), and returned to normal (9·2–10·2 mg/100 ml) when the skin improved. During a typical exacerbation (1962), the plasma protein concentration was 6·3 g/100 ml, albumin 3·0 g/100 ml, alkaline phosphatase up to 40 K-A units, ultra-filtrable calcium 4·4 mg/100 ml. Fæcal fat excretion was 19·9 g/day (over five days, on a normal ward intake). Fig 1 summarizes some of the biochemical data during one admission; changes in body temperature reflected changes in the skin condition.

Like the serum calcium, the alkaline phosphatase, plasma proteins and fæcal fat excretion became normal when the skin improved.

Other investigations: Blood urea 26 mg/100 ml, creatinine clearance 125 ml/min, plasma phosphorus 2·9–5·1 mg/100 ml, theoretical renal phosphate threshold 3·6 mg/100 ml (normal), bilirubin 0·2 mg/100 ml, thymol turbidity 3, SGOT 23 units/100 ml. Glucose tolerance (during remission) normal. Histidine loading test – a gross excess of formiminoglutamic acid in the urine. Skin loss – shed skin was collected from the whole body over a five-day period. The average weight was approximately 30 g/day, containing 54 mg calcium/day and 15·5 g protein/day.

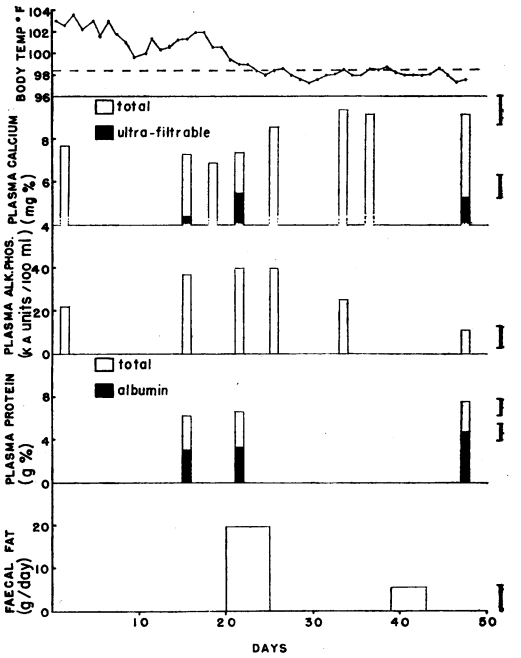

Fig 1 *Some biochemical data during an exacerbation of pustular psoriasis. The normal ranges for each estimation are indicated by the bars on the right hand side*

X-rays of hands, feet, spine and skull showed no abnormality apart from osteoarthritic changes.

Small bowel meal normal. Jejunal biopsy (during remission) normal.

Comment

Approximately 40% of the total serum calcium is protein bound and biologically inactive. Low plasma protein concentrations reduce the protein-bound calcium; this may be sufficient to cause a low total serum calcium (Fowler *et al.* 1961). Although the hypocalcæmia in this patient has never been severe enough to cause tetany, and although his plasma proteins do fall, the initially low ultra-filtrable calcium concentration shows that the ionized calcium concentration is reduced. The ultra-filtrable calcium has been found to be low during three other admissions with generalized pustular psoriasis. Also, the plasma alkaline phosphatase is raised during the period of hypocalcæmia and returns to normal a few days after the serum calcium becomes normal. Serum bilirubin, thymol turbidity, and SGOT were normal when the alkaline phosphatase was raised. Further investigation is planned to decide if bone is the source of the raised alkaline phosphatase.

Four possible causes of hypocalcæmia were considered. Loss of calcium in the skin, though

much greater than normal, is too small to be a significant cause. Hypoparathyroidism is thought unlikely as the plasma phosphorus and theoretical renal phosphate threshold were normal and the alkaline phosphatase raised. Normal blood urea and creatinine clearance exclude renal failure as a cause of hypocalcæmia. Fæcal fat excretion was increased, however, on two occasions during an exacerbation of pustular psoriasis, and malabsorption of calcium and vitamin D seems the most likely explanation of the hypocalcæmia. The association between steatorrhœa and exfoliative skin disease was reviewed by Wells (1962).

Several observers have reported that hypocalcæmia may precipitate various skin diseases, e.g. impetigo herpetiformis (Beek 1961), eczema and exfoliative dermatitis (Dent & Garretts 1960), and psoriasis (Vickers & Sneddon 1963, Montgomery 1964). It was originally thought that this patient had intermittent steatorrhœa causing hypocalcæmia, which produced an exacerbation of the pustular psoriasis.

However, in this patient fæcal fat excretion has been normal during three remissions of the pustular psoriasis. No obvious primary cause of steatorrhœa has been found. There is no clinical evidence of pancreatitis or liver disease, though these cannot yet be excluded definitely. The normal jejunal biopsy, although taken when the pustular psoriasis was in remission, is thought to make a gluten sensitivity unlikely. It is therefore interesting to speculate on the possibility that an exfoliative skin disease could itself produce malabsorption. Hindle & Creamer (1965) recently found 3 patients in whom a history of dermatitis preceded evidence of steatorrhœa and suggested the possibility that the skin disease might be the cause of the steatorrhœa.

If this patient relapses, we are hoping to do further investigations including a metabolic balance, pancreatic enzyme measurements and, if possible, a jejunal biopsy when there is malabsorption, to study further the relationship between skin and gut.

REFERENCES
Beek C H (1951) *Dermatologica* **102**, 145
Dent C E & Garretts M (1960) *Lancet* i, 142
Fowler D I, Fore D J & Cooke W T (1961) *Lancet* ii, 284
Hindle W & Creamer B (1965) *Brit. med. J.* (in press)
Montgomery P R (1964) *Proc. R. Soc. Med.* **57**, 1128
Vickers H R & Sneddon I B (1963) *Brit. J. Derm.* **75**, 419
Wells G C (1962) *Brit. med. J.* ii, 937

Dr E J Moynahan: Has gluten-free diet been tried?

Dr G C Wells: Dr Bold's work suggests that on occasion a widespread skin disease might induce a transient state of intestinal malabsorption. Certainly this patient had steatorrhœa and a positive Figlu test during an exacerbation of his pustular-erythematous psoriasis. During remission all tests for malabsorption have been negative and jejunal biopsy showed no abnormality. We have not, therefore, given him a gluten-free diet.

We have not got a very satisfactory explanation for the folic acid deficiency which is not rare in eczema and psoriasis (Knowles *et al.*, 1963, Lancet i, 1138), and I think that this idea of transient malabsorption during exacerbations of skin disease deserves attention.

Dr L Fry: So-called 'adult cœliacs', when in a state of clinical remission, may have no steatorrhœa, but steatorrhœa can be precipitated by any illness in these subjects. Is this happening in this case? Small intestinal biopsy and appropriate tests on the mucosal specimen are necessary for establishing the diagnosis, or possible other causes for malabsorption.

Dr M Garretts: We are used to seeing patients with phases of malabsorption, without infection being present.

It is interesting to consider psoriasis as a possible malabsorption disease. Dr MacKenna has reminded us from time to time that the increased pigmentation around healing lesions of psoriasis requires explanation. We do not really know the mechanism of pigmentation associated with steatorrhœa.

Postscript (14.4.65): Jejunal biopsy during recent attack showed definite flattened mucosa. – P W M C.

Porphyria Cutanea Tarda presenting as 'Neurotic Excoriations'

W E Beer MB MRCPEd (for H J Wallace FRCP)

J B, female, aged 34

History: At the age of 17 the patient developed a red irritating rash on her face and upper limbs. Since then she has never been entirely free of trouble. There was a relapse of the condition during August 1964 while she was on holiday in Majorca (she had, in the past, been on several occasions to the Mediterranean area but had had no such trouble). She had never observed any seasonal variation in her condition.

On examination (24.9.64): Numerous excoriations on the face and upper limbs interspersed with many scars. A few lesions were also present on the lower limbs (Figs 1 and 2). No abnormalities detected in other systems.

The patient made vague mention of having had blisters and added that her skin had always been easily vulnerable. This was thought to be a facile explanation for her self-inflicted injuries. A diagnosis of 'neurotic excoriations' was made, but with some reservations as her general demeanour and the presence of lesions on the fingers were thought not to be entirely consistent.