Acknowledgments: I am grateful to the consultants who have allowed me to give details of their patients. A fuller account of this syndrome and relevant literature will be given elsewhere. Some illustrations can be found in the references given and others will appear in the fuller account (Mallinson *et al.* 1974).

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Dr R E Church: While bizarre widespread eruptions may occur in association with underlying neoplasm such as carcinoma of the bronchus I think the clinical appearance of the rash which we are discussing is unique. Nevertheless it may vary in its appearance from time to time and in making this diagnosis I do not think we should put too much stress on the rash alone but should also take into account other features of this syndrome such as the sore red tongue, diabetes, weight loss and anæmia which Dr W A F Crane and I described in our case and which have been present in those cases which have come to light since.

Dr R R Harman: In his description of this group of cases Dr Warin has not mentioned the very odd features on the ear lobes in Case 2 and also in Case 6. Dr R P Warin's patient had large perforations of the lobes due to the pressure of clip-on ear rings, and the other patient had a large part of the left ear lobe missing; the patient attributed this to pressure from a hair-net rubber band. All cases have had very thin skin tending to break down in pressure areas and these unique ear signs may indicate an important dermal thinning and fragility.

Was there no comment on the ear lobes in the other cases?

Dr R D Sweet: Perhaps the most remarkable thing about my patient (Case 1) was the rapid disappearance of the rash after the pancreatic tumour had been removed. The patient's tongue was normal in 24 hours and her extensive eruption cleared within a week. Attempts to reproduce this eruption with a commercial glucagon preparation with patch tests and intradermal injections were unsuccessful.

Dr R Summerly: As far as I am aware, the anti-pancreatic lipase activity of the tetracyclines has only been established in the *in vitro* situation. The beneficial effect, reported by some clinicians, of systemic tetracyclines on the necrolytic erythema of this syndrome may be due to such inhibition of the pancreatic lipase and should be tested in the *in vivo* situation.

Alopecia Areata, Vitiligo, Scleroderma and Ulcerative Colitis

D M Thompson MB MRCP
(for T W E Robinson MB MRCP
and J Lennard-Jones MD FRCP)
(*University College Hospital, London WC1E 6JJ*)

V F, man aged 31. Born in Malta

History: 1968: attended London Hospital with a subtotal alopecia. Since then hair has regrown and fallen out twice. 1969: developed soreness of mouth. Eroded and blistered areas noted on palate and anus. Later appearances highly suggestive of lichen planus. 1970: lichen simplex chronicus of trunk and legs. 1971: attended Westminster Hospital with repeated episodes of nonspecific urethritis. 1972: attended UCH with a further episode of urethritis associated with painful and stiff knees, ankles, wrists, fingers, elbows and lumbar spine. X-rays showed calcaneal spurs and diagnosis of incomplete Reiter's syndrome made. July 1973: seen at UCH because of pruritus of legs. White patches on back present for previous three months. Recent lower abdominal pain with loose stools in morning. Similar episode for one year, four years ago. Since then diarrhœa has become progressively worse, with passage of 8 stools during day and 2 at night. No blood or mucus.

Family history: His brother has thyroid trouble and his mother has diabetes.

On examination: Almost total hair loss (Fig 1A). Vitiligo of back (Fig 1B). Sclerodermatous pigmented areas on arms and lower legs (Fig 1C). Extending ivory coloured wrinkled discoid areas on buttocks. Exophthalmos, gynæcomastia, normal genitalia.

Investigations

1972: Hb 13.2 g/100 ml. WBC 7200/mm³ (neutros. 72%, lymphos. 20%, monos. 6%, eosinos. 13%). Persistant eosinophilia of up to 2400 cells/mm³. ESR 40 mm in first hour (Westergren). X-rays of lumbar spine, sacroiliac joints, knees and chest normal. X-rays of heels showed calcaneal spurs.

1973: Hb 15.2 g/100 ml. WBC 5800/mm³ (neutros. 72%, lymphos. 20%, eosinos. 4%). ESR 10 mm in 1 h (Westergren). Antinuclear factor positive in titre of 1/160. No thyroid, gastric, smooth muscle and mitochondrial antibodies detected.

Proteins: Albumin 4.5, globulin 3.9, total 8.4 g/100 ml. IgA 230, IgM 25, IgG 1920 mg/100 ml. No cryoglobulin detected.

Lymphocyte marker tests (Dr M Greaves): T lymphocyte markers: E (sheep) rosettes 78.3% reactive cells (normal). B lymphocyte markers: anti-immunoglobulin 6.0% reactive cells (very low). Phytohæmagglutinin response of lymphocytes normal.

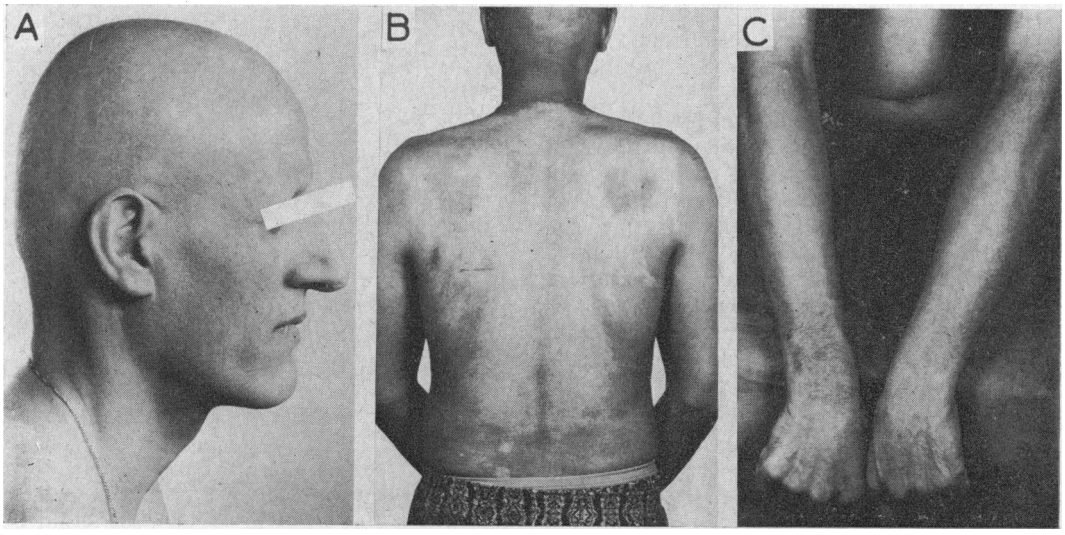

Fig 1A, *alopecia subtotalis.* B, *vitiligo and scleroderma on back.* C, *sclerodermatous, pigmented changes on forearms*

Normal investigations: serum urea, electrolytes, creatinine glucose, uric acid, SGOT, calcium, phosphorus, alkaline phosphatase, bilirubin, protein-bound iodine, creatinine clearance, urinary indican, fæcal culture, fæcal fats, ECG, respiratory function tests including diffusion carbon monoxide, Rose Waaler and latex fixation test, rapid plasma reagin test and Reiter protein complement fixation test.

Radiology: Chest, hands, barium swallow and meal normal. Barium enema (Fig 2): whole of colon uniformly narrowed with irregular ulceration of the mucosa and complete loss of haustral pattern; 'backwash ilietis' causing dilation of terminal ileum; rectum normal. The appearances were those of a pericolitis (probably ulcerative colitis).

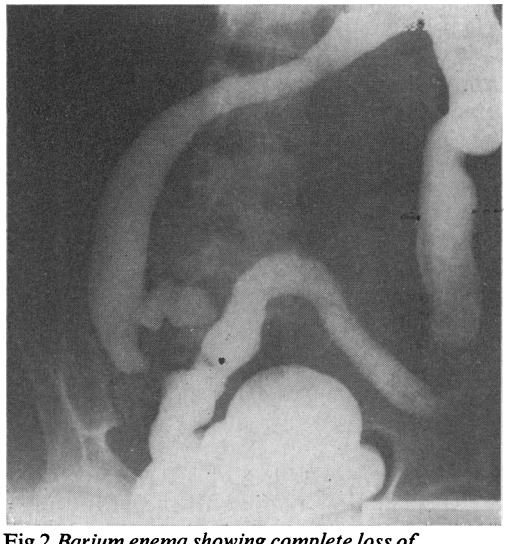

Fig 2 *Barium enema showing complete loss of haustration in colon with normal rectum*

Special investigations: Proctoscopy showed a normal rectum but sigmoidoscopy (Dr T Warnes) revealed ulcerative colitis. Colonoscopy (Dr C Williams) showed moderately active total colitis with rectal sparing; slough-covered patches and ulcers is discending colon and transverse colon; large polyp at splenic flexure.

Histopathology: Mouth (London Hospital): report suggested possibility of lichen planus, chronic lupus erythematosus or benign mucous membrane pemphigoid. Rectum: normal mucosa. Leg (Dr A Jarrett): condensation of collagen and loss of skin appendages; epidermis somewhat thinned and there is increased pigmentation of the basal layer. Buttock shows similar changes; epidermis more atrophic and almost all appendages except a few sweat glands have disappeared from the dermis.

Treatment: Sulphasalazine and prednisolone enemata.

Comment

The association of lichen planus with alopecia areata, vitiligo and ulcerative colitis has been recently described (Tan 1974). Alopecia areata has been noted in patients with ulcerative colitis (Müller & Winkelmann 1963) but there is only one reported case of a patient with both ulcerative colitis and scleroderma (Bicks *et al.* 1958). In this patient, symptoms of ulcerative colitis preceded her other abnormalities by five years. Her sclerodermatous changes were confined to skin and lungs and there was no involvement of other parts of the gastrointestinal tract but symptoms

of wide spectrum of connective tissue disease were a feature of her illness. Examination of her colon at post-mortem showed histological features of both diseases.

The colon is relatively infrequently affected in scleroderma. Meszares (1959) found involvement of the colon in only 2 out of 50 patients with systemic sclerosis examined by barium enema. In his review of these patients and 13 others the radiological abnormalities noted were wide mouthed sacculations alternating with narrowed areas and in all cases the œsophagus was also involved. It seems therefore almost certain that, despite the unusual feature of sparing of the rectum our patient has ulcerative colitis. The significance of the low B cell population is as yet uncertain.

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Dr P F Borrie: I think we must be careful about the word scleroderma. Large bowel involvement is well recognized in systemic sclerosis. The present case is, however, obviously not one of systemic sclerosis. I would like to suggest that the cutaneous lesion is lichen sclerosus et atrophicus and, further, that this is responsible for the depigmentation and that there is no vitiligo.

Malignant Angioendothelioma of the Scalp

Anthony du Vivier[1] MRCP (for M Feiwel MD FRCP)
(St Mary's Hospital, Praed Street, London W2)

J H, man aged 77

History: Presented with a two-month history of a lesion on his right forehead which was rapidly increasing in size. No other complaints.

On examination: A purple and in some areas black tumid lesion on the right frontal region of the scalp surrounded by a flat 'port-wine'-like discolouration extending over the forehead.

Investigations: Full blood count, ESR, liver function tests, chest and skull X-rays normal.

Skin histology: The dermis showed vascular spaces lined by malignant endothelial cells. Also scanty solid clumps of atypical cells with large vesicular nuclei, foci of lymphoid cells and of hæmorrhage. The appearances were those of an angioendothelioma towards the less malignant end of the range (Dr J K Blenkinsopp).

[1]Present address: Department of Dermotology, Scripps Clinic, 476 Prospect Street, La Jolla, California 92037 USA

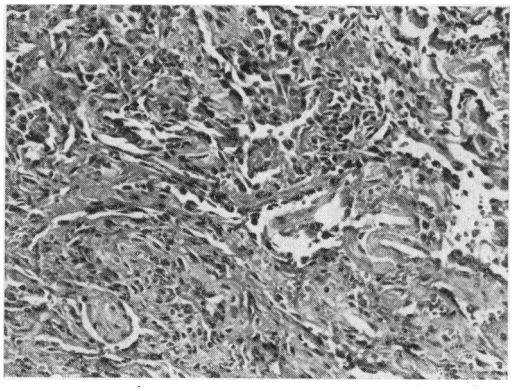

Fig 1 *Shows the histology of the lesion. HE× 120*

Comment

This patient's skin tumour conforms well in appearance and histology to the description by Wilson Jones (1964) of these rare lesions. The prognosis is not good and the lesions are difficult to treat. This tumour was too extensive for surgery and was recommended for radiotherapy.

REFERENCE
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The following cases were also presented:

Follicular Mucinosis
Dr H M Barnes (for Dr I Sarkany)
Xeroderma Pigmentosum with Neurological Abnormalities
Dr I Sarkany
Case for Diagnosis: Probably Pyoderma Gangrenosum
Dr T W E Robinson & Dr D A Birkett
Cœliac Disease with Dermatitis Herpetiformis
Dr D Thomson (for Dr T W E Robinson)
Cœliac Disease with Dermatitis Herpetiformis
Dr D Thomson (for Dr C M Ridley)
Lichen Planus, Poikiloderma and ? Lupus Erythematosus
Dr D Thomson (for Dr T W E Robinson)
Nodular Livedo Reticularis with Ulcerative Colitis
Dr P M Hudson
(for Dr T W E Robinson & Dr J E Lennard-Jones)
Cold Urticaria
Dr C A Ramsay
Relapsing Polychondritis
Dr H J N Bethell (for Dr E J Moynahan)

(Meeting to be continued)