Self-Assessment Colour Review

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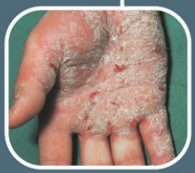
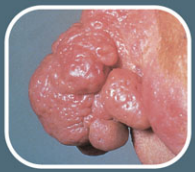
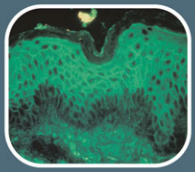
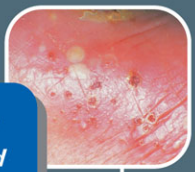




Dermatology



Ronald Marks



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Dermatology

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Preface

Dermatology and ophthalmology are reputedly the two specialties in medicine that have the 'most diseases'. If this is the case, it may be because in only these two specialties, compared to all the others, the disorder is obvious. This makes dermatology a special skill, tending to attract the 'Sherlock Holmes'-type of physician. This kind of individual will, in clinical practice, assiduously look for clues and will experience genuine pleasure when involved in the deductive process. This book should satisfy his/her intellectual lust.

The book has additional other important purposes. It has been recognized since the 'beginning of time' that there is no better way of learning than being tested time and time again. The collection of questions and answers in this book covers most aspects of clinical dermatology and once its content has been mastered the reader will have made an important start to being proficient at sorting out (recognizing) skin disorders.

Ronald Marks

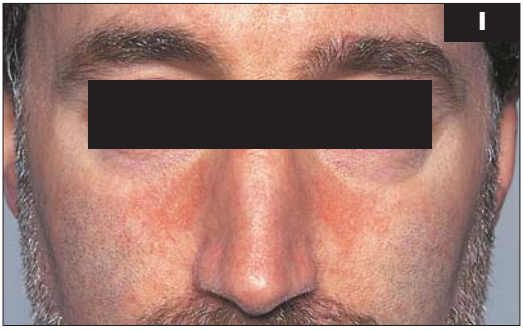
Acknowledgements

I gratefully acknowledge the assistance of Dr Richard Goodwin in assembling the illustrations.

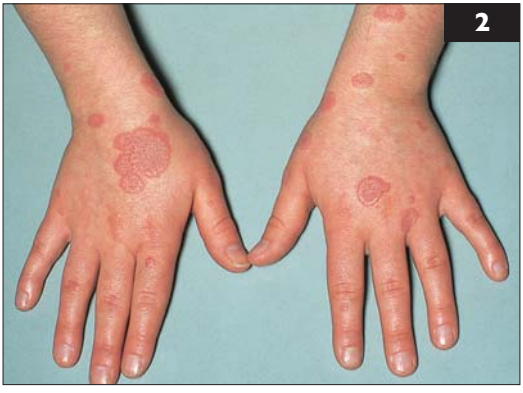
I am also extremely grateful to the Department of Medical Illustration of University Hospital of Wales for taking many of the photographs.

Abbreviations

AA	alopecia areata
CDLE	chronic discoid lupus erythematosus
CDNCH	chondrodermatitis nodularis chronica helicis
DM	dermatomyositis
DSAP	disseminated superficial actinic porokeratosis
EB	epidermolyis bullosa
EBA	epidermolyis bullosa acquisita
EM	erythema multiforme
EMPD	extramammary Paget's disease
EN	erythema nodosum
EPS	elastosis perforans serpiginosa
GA	granuloma annulare
HIV	human immunodeficiency virus
HPV	human papillomavirus
HSP	Henoch–Schönlein purpura
IgG	immunoglobulin G
JPD	juvenile plantar dermatosis
NLD	necrobiosis lipoidica diabeticorum
NME	necrolytic migratory erythema
PASI	psoriasis area severity index
PCT	porphyria cutanea tarda
P–J	Peutz–Jeghers disease
PLE	polymorphic light eruption
PLEVA	pityriasis lichenoides et varioliformis acuta
PRP	pityriasis rubra pilaris
PTM	pretibial myxoedema
PUVA	photochemotherapy with ultraviolet A
SLE	systemic lupus erythematosus
SSGH	senile sebaceous gland hyperplasia
TNF-α	tumour necrosis factor alpha
UV	ultraviolet
UVR	ultraviolet radiation



1 A male complains of increasingly severe dandruff and an itchy rash on the face (**1**). No other area is involved. He is in other respects well and there is no significant history of previous disease. He is prescribed a steroid cream, which provides only temporary relief.
i. What is the most likely diagnosis?
ii. What differential diagnosis should be entertained?
iii. What investigations should be undertaken?

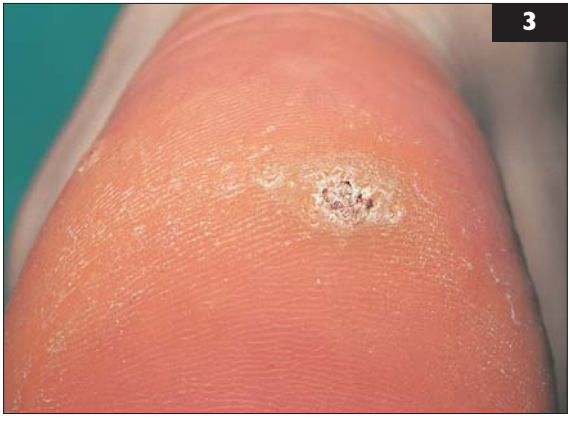


2 A 52-year-old female became ill while on holiday in Kenya. At first there was a faint rash on her trunk. The following day the eruption was very much worse with large deep red blotches all over her skin. She also felt unwell and had a headache and fever. On the third day she was much worse, with the skin showing blistering in places and peeling in others (**2**). The oral and genital mucosae had also developed erosions.
i. What is the most likely diagnosis? How does it differ from other similar disorders?
ii. What investigations may help in establishing the diagnosis?
iii. Discuss the management.

1, 2: Answers

1 i. The most likely diagnosis is seborrhoeic dermatitis.
ii. The differential diagnosis should include psoriasis, allergic contact dermatitis to a topical application, and atopic dermatitis, although this latter would be unusual occurring for the first time in an adult. Psoriasis is not usually itchy, but certainly can be. Other disorders such as rosacea and lupus erythematosus are seldom itchy.
iii. If there is doubt concerning the clinical diagnosis, it is worth performing a biopsy as although an indeterminate result may not be helpful, a positive result would be conclusive. If a 3 or 4 mm punch biopsy is performed without sutures being inserted, scarring will not be a problem. Patch testing to cosmetics or medicaments will be appropriate if allergic contact dermatitis is suspected. If the patient has severe seborrhoeic dermatitis occurring for the first time, the possibility of human immunodeficiency virus (HIV) disease or other form of immunosuppression should be considered and the appropriate blood tests performed.

2 i. The generalized nature, the severity, and the rapid progression all favour a drug reaction. The picture is more like that of toxic epidermal necolysis than severe erythema multiforme (EM), although there may be overlap in some patients. In EM the mucosal involvement is earlier and more severe and the rash tends to remain in distinct elements rather than becoming generalized. On investigation it was found that she had been taking mefloquine as malaria prophylaxis and this is a rare but recognized cause of toxic epidermal necrolysis in a few subjects.
ii. Skin biopsy showed severe necrotic change in the upper epidermis, with a dense inflammatory cell infiltrate subepidermally and invading the epidermis.
iii. The patient was evacuated by air ambulance back to the UK and was admitted to an intensive care unit. She was treated as though she had severe burns because of the severe dehydration and susceptibility to infection as a result of the extensive damage to the skin barrier. She was also treated with prednisone 60 mg/day. This patient survived, but must be counted as fortunate as there is a mortality rate of approximately 50%.



3 i. What is the lesion in **3**?
ii. How should it influence its owner's sporting activities?
iii. Discuss the treatments for this condition.

4 A 70-year-old female, who has lived in Florida for the past 30 years, presents with an increasing skin problem on both lower legs. Close examination shows that there are numerous small warty lesions on the fronts of the legs and many brownish macules. She also has many small distinctive round warty patches, which are surmounted by a horny ridge and seem to have an atrophic centre. In addition, there is a crusted plaque (2 × 1.5 cm) in the middle of the left shin.
i. What is the diagnosis?
ii. What other causative agencies have been blamed for the distinctive lesions that she has?
iii. What is the likely diagnosis of the crusted lesion?

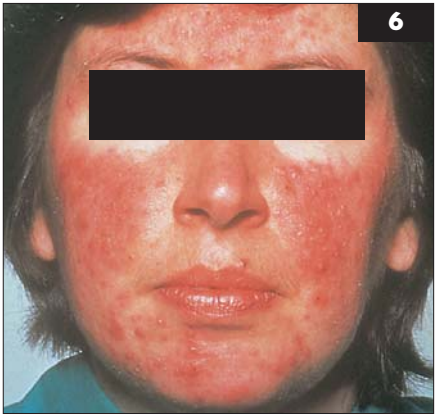
5 A patient, who works on a farm, has had a tender red nodule on the dorsum of his right index finger for the last 3 days.
i. What is the most likely diagnosis?
ii. How did he become infected?
iii. Are there any consequences that could be expected?

3–5: Answers

3 i. The lesion is a plantar wart. Callosities are found at sites of friction, and are not as well marginated. Corns are mostly found on the tarsal arch and do not have tiny black dots on the surface as do plantar warts.
ii. It is customary to recommend that the sufferer avoid swimming because of the risk of spreading wart infection but it is doubtful whether this reduces spread. Plantar warts are quite tender on pressure and some children use this as an excuse to avoid sporting activities, but they should be encouraged to do as much as possible.
iii. In general, treatment is not very effective and is often quite painful. In addition, all plantar warts eventually resolve spontaneously. For these reasons some dermatologists recommend reassurance without active treatment. Active treatments are all destructive and include freezing with liquid nitrogen or solid carbon dioxide (cryotherapy), surgical removal by curettage and cautery, ablation by laser beam, or chemotherapy with keratolytics (high concentrations of salicylic acid) or antimitotic agents, such as podophyllin extracts or bleomycin.

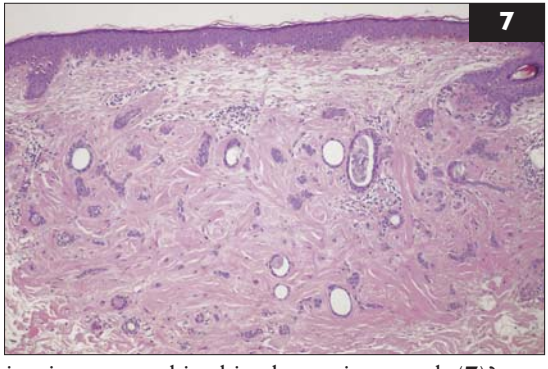
4 i. The diagnosis is almost certainly chronic solar damage with numerous solar keratoses, solar lentigines, and lesions of disseminated superficial actinic porokeratosis (DSAP).
ii. Lesions of porokeratosis have been blamed on immunosuppression, genetic factors, and solar exposure.
iii. The crusted lesion is almost certainly either a squamous cell carcinoma or a basal cell carcinoma – both of which are more frequent in sun-damaged sites and where there are lesions of porokeratosis.

5 i. The most likely diagnosis is orf. This is an infection caused by a DNA virus of the poxvirus group, which spreads to man from sheep and goats.
ii. The hands and fingers are sites commonly involved as the infection is transmitted by contact with affected sites.
iii. The condition usually lasts 10–14 days and heals without scarring. However, in a significant proportion of affected individuals an attack of erythema multiforme follows some 2–4 weeks later.



6 A female was 24 years of age when she first noticed that she was blushing much more frequently than she had done previously and that the flushes lasted for longer than they used to. Not only did she blush at minor embarrassments, but she flushed deeply with spicy food and alcoholic drinks. Now, aged 29, she presents because of the persistent redness of her cheeks, chin, and forehead, and the 'pimples' that have appeared in the red areas (**6**). These seem to come in crops and are sometimes accompanied by pustules.

i. What is the differential diagnosis?
ii. What investigations will help establish a diagnosis?
iii. What are the treatment options for the most likely diagnosis?



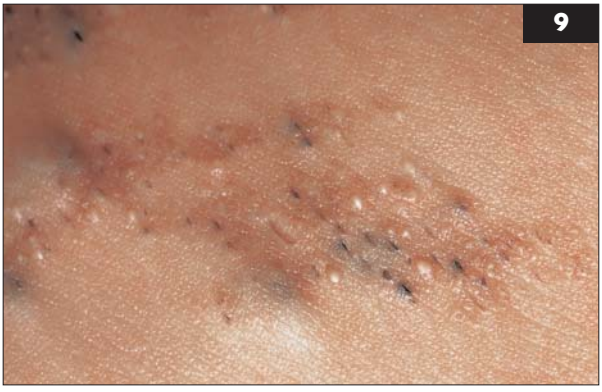
7 i. What condition is presented in this photomicrograph (7)?
ii. What clinical features develop in this condition?
iii. What treatment should be recommended?

6, 7: Answers

6 i. The most likely diagnosis is rosacea, but the differential diagnosis includes seborrhoeic dermatitis, although this disorder is scaly (rosacea is not) and also affects the scalp and facial flexures. In contrast, rosacea mostly affects the facial convexities. The differential diagnosis also includes systemic lupus erythematosus (SLE) and dermatomyositis (DM). The former is a systemic disorder which is accompanied by multiple positive laboratory findings including the presence of anti-DNA antibodies, anaemia, leucopenia, and thrombocytopenia. DM is distinguished by the facial rash having a mauve–lilac hue (heliotrope) and occurring mainly on the upper face as well as the back of the hands. Limb girdle muscular weakness and tenderness accompany the skin disorder. Neither of these conditions causes papules or pustules to appear on affected skin.
ii. A full blood picture, a test for anti-DNA antibodies, and a blood test for muscle enzymes would help exclude SLE and DM. A skin biopsy would distinguish between SLE and rosacea as in the former there is tight 'cuffing' of the small blood vessels with lymphocytes and degenerative changes in the basal epidermal cells. Direct immunofluorescence tests would detect deposits of immunoglobulin G (IgG) and complement component C3 at the dermoepidermal junction in lupus erythematosus.
iii. The treatment of choice for acute rosacea is oral tetracycline. It has been suggested that it is the anti-inflammatory effect rather than any antibacterial action that is responsible for improvement. All the tetracyclines reduce the number of papules and calm the inflammation within 3–4 weeks. A low dose of doxycycline can be administered once or twice daily and is very well tolerated (50 mg twice daily). Oral erythromycin is also effective. Topical metronidazole gel (0.75%) used twice daily is quite effective as is topical azelaic acid cream (20%). Pulsed dye laser treatment has been shown to improve the redness and flushing considerably.

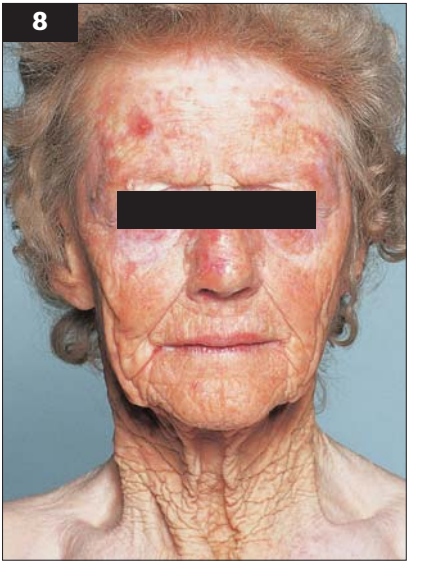
7 i. The condition is the 'naevoid disorder' of sweat gland elements known as syringoma. The histological picture must be carefully distinguished from morphoeic basal cell carcinoma and the rarer condition of desmoplastic trichoepithelioma.
ii. Characteristically small (1–2 mm) pink papules occur on the lower eyelids and on the upper cheeks. In some cases very large numbers of papules also occur over the trunk and limbs. They may do so over relatively short periods of time and the condition is then known as 'eruptive hidradenoma'.
iii. Unfortunately, the only available treatment is the ablation of individual lesions, for example by laser ablation.

8 Severe sunburn affecting the face, upper trunk, and arms is unlikely to occur in northern Europe.
i. Comment on this statement.
ii. Briefly discuss the aetiopathogenesis and pathology of sunburn.
iii. What may be the long-term sequelae of repeated or chronic sunburn?



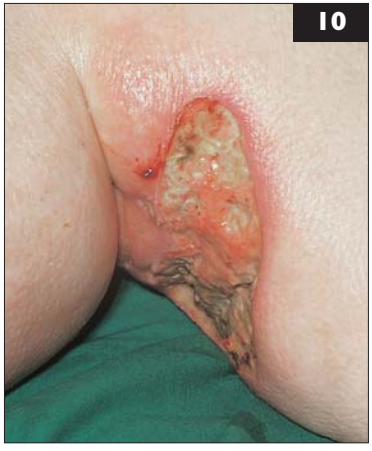
9 A 30-year-old male has been aware of what he called a birth mark on the left side of his abdomen since early childhood (**9**). In the past few years occasional inflamed spots have developed in the affected area.
i. What is this condition?
ii. Why do inflamed spots develop on the lesion?
iii. Name related naevoid conditions.

8, 9: Answers



8 i. Sunburn can certainly occur in northern Europe depending on the time of year, the length of the exposure, and the vulnerability of the subject. The vulnerability of the subject is primarily a function of the depth of his or her pigmentation.
ii. Sunburn is primarily due to a critical dose of solar ultraviolet radiation (UVR) – in particular UVR in the medium waveband (known as UVB) of 190–220 nm band width. The UVB penetrates the epidermis but does not penetrate into the dermis. The damage caused to the epidermal cells results in cell death (sunburn cells), spongiosis, subepidermal blistering, and general inflammatory change. The process is complex, involving activation of the prostaglandin cascade and DNA damage with the formation of thymidine dimers.
iii. Because damage from solar exposure does not cause dermal destruction, scarring and keloid formation do not result. Similarly, permanent alteration in pigmentation does not occur. However, exposure to UVR in the long term causes both melanoma and nonmelanoma skin cancer. The present day epidemic of melanoma and the increase in incidence of basal cell carcinoma, solar keratosis, and squamous cell carcinoma is due mostly to the increased annual dose of UVR. Apart from these important results of 'chronic burning', the altered appearance of exposed skin with its lines and wrinkles and yellowish discolouration, known as solar elastosis, causes considerable cosmetic disability (**8**).

9 i. This is a comedone naevus, a type of epidermal hamartoma in which there are many follicular canals packed with horny debris (as in ordinary blackheads).
ii. The inflamed spots are analogous to the papules or papulopustules in acne and derive from the comedones.
iii. Related naevoid conditions include warty epidermal naevus, naevus sebaceous, 'organoid naevus'.

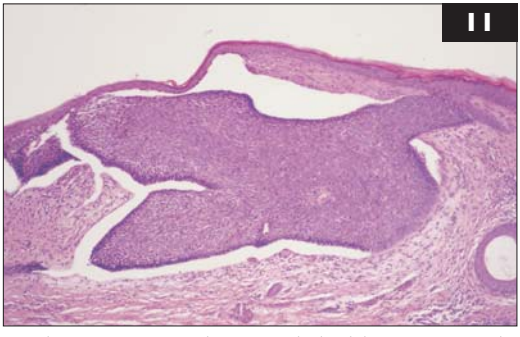


10 A 27-year-old male has had attacks of diarrhoea with blood and mucus since the age of 18. He appeared otherwise quite well and seemed to respond to oral sulfasalazine. During a flare-up of his bowel condition last summer he developed some inflamed papules and pustules on the lower legs. These lesions looked like acne, but one spot became larger than the rest and developed into an ulcerated area of 2 cm^2 (**10**). Despite careful treatment with nonadherent dressings and antibiotics, the open area spread within days to produce an ulcer 4 cm^2 in area. It was shallow and had mauve–blue, slightly undermined edges. Bacteriological swabs grew a mixed bacterial flora; sometimes *Pseudomonas* spp. was recovered, but the lesion did not respond to topical mupirocin.

i. What is the differential diagnosis?

ii. Discuss the pathogenesis of the most likely diagnosis.

iii. What treatments are available for the most likely diagnosis?



11 A 34-year-old male presents with several darkly pigmented spots on his face, chest, and arms. One of the lesions is removed under local anaesthetic and the pathology is shown in **11**. The patient's 12-year-old daughter also has some pigmented spots on her face and chest.

i. What is the diagnosis and why?

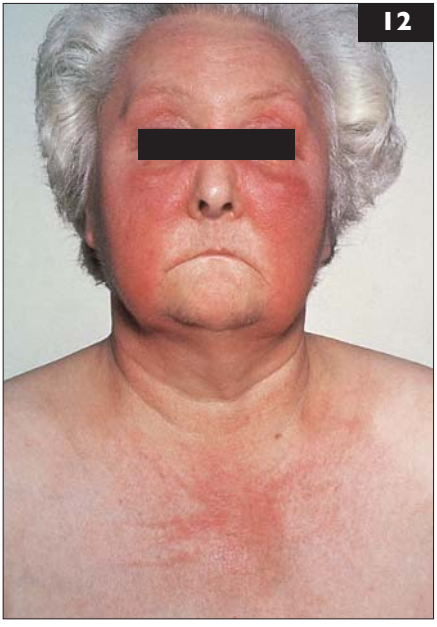
ii. Are there any accompanying stigmata or any bone abnormalities that are characteristic of this condition?

iii. Discuss the management of this condition.

10, 11: Answers

10 i. The diagnosis is almost certainly pyoderma gangrenosum. This condition occurs in the course of ulcerative colitis and other chronic inflammatory disorders including Crohn's disease and rheumatoid arthritis. It also occurs in paraproteinaemias and leukaemic disorders. In a proportion of cases (perhaps 30%) no underlying cause can be found. It can spread with frightening rapidity, but can also heal quickly. Other ulcerative conditions that should be considered include a form of vasculitis (e.g. polyarteritis nodosa), and a skin infection such as a streptococcal condition or infection with a *Mycobacterium* spp.
ii. Pyoderma gangrenosum is not due to a skin infection. It has been thought to be due to a small vessel vasculitis, but skin biopsies usually do not confirm the presence of this. It is generally assumed that an immunological reaction of some type is involved leading to acute ischaemic ulceration and subsequently infection.
iii. Various treatments have been used and reported as successful including the low-dose tetracyclines minocycline and doxycycline, systemic corticosteroids, dapsone, clofazimine, mycophenolate mofetil, and ciclosporin, the latter drug being the most frequently used. Recently, biological immunomodulators such as infliximab have been used successfully.

11 i. This patient is suffering from the basal cell naevus syndrome (Gorlin's syndrome). The development of multiple pigmented lesions, which turn out to be basal cell carcinomas, in a relatively young individual is quite typical. The development of similar lesions in the patient's daughter is also consistent as the condition is inherited as a Mendelian dominant characteristic.
ii. Minute pits on the palmar skin are a common accompaniment. There may be five or more of these, each less than 1 mm in diameter. Bony abnormalities may occur including cysts in the mandible and bifid ribs. Other abnormalities include frontoparietal bossing of the skull giving a relative macrocephaly and various vertebral abnormalities.
iii. Individual lesions should be removed as they appear. However, excision may become impractical when very large numbers develop and for these patients the use of oral retinoids has proved invaluable. Both isotretinoin and acitretin have been most frequently used and regular administration of one of these greatly reduces the rate of appearance of new lesions. Unfortunately, the regular administration of one of these inevitably causes side-effects (dry lips, loss of hair, dry itchy skin), which makes compliance difficult.



12 A female has developed a rash on her face in the previous 6 weeks and complains that it is gradually worsening (**12**). She also feels unwell, tired, and weak, and the rash is spreading to other areas of skin. It is particularly difficult to comb her hair and to sit up from the horizontal position. She finds that her shoulders and thighs are quite tender to the touch.
i. What is the diagnosis?
ii. What laboratory tests should be performed to confirm the diagnosis?
iii. What other areas of the skin may be affected?
iv. What diseases may be associated with this disorder?
v. Outline a management strategy for patients affected by this disease.

13 An overweight 64-year-old male presented with a painful, tender red nodule that had developed on the right side of the upper part of the back at the site of a symptomless swelling that he had noted over the past few months. He was prescribed antibiotics, but the condition did not improve. After 3 weeks the centre of the nodule broke down and drained blood-stained yellowish viscid pus.
i. What is the differential diagnosis and what is the most likely diagnosis?
ii. Discuss the pathogenesis and the pathology.
iii. Comment on the management for this condition.