Multiple facial plaques
A case study

Case study
Mrs AA, female, 27 years of age and of Middle Eastern descent, presented with a 1 year history of nonpruritic facial plaques. On examination, these plaques were 0.2–1.0 cm, scaly, atrophic, annular, hyperpigmented and located on her forehead, nose, chin and adjacent to her lips (Figure 1). She was otherwise well apart from iron deficiency anaemia treated with ferrous sulphate and recurrent headaches treated with paracetamol. She had completed a course of cephalexin 2 months before for a urinary tract infection. She had not taken any other medications in the preceding 12 months. Mrs AA had no family history of skin disorders.

Question 1
What differential diagnoses would you consider?

Question 2
What further history would you take?

Question 3
What areas would you examine?

Question 4
What investigations would you perform?

Case study continued
Histopathological examination of a punch biopsy from Mrs AA’s cheek revealed atrophic epidermis, basal cell vacuolisation, scattered Civatte bodies, marked lymphocytic infiltrate and pigmented incontinence. On immunofluorescence there were numerous cytoid bodies that stained with immunoglobulin A (IgA) and immunoglobulin M (IgM). A broad band of fibrinogen along the dermal-epidermal junction was present, but no deposition of immunoglobulin or complement could be detected. The report stated that the histopathological findings were consistent with actinic lichen planus or an incomplete picture of discoid lupus erythematosus. Blood tests were normal except for: haemoglobin 102 g/L, erythrocyte sedimentation rate (ESR) 35 mm/hr, and antinuclear antibody (ANA) 1:160 (extractable nuclear antibodies [ENA] and anti-double stranded DNA [dsDNA] negative).

Question 5
What is the likely diagnosis?

Question 6
What is the treatment for this condition?
**Answer 1**

The differential diagnosis of this presentation includes discoid lupus erythematosus (DLE), melasma, granuloma annulare, polymorphous light eruption, secondary syphilis, sarcoidosis, fixed drug eruption, erythema dyschronicum perstans, morphea and actinic lichen planus (Table 1).1

**Table 1. Differential diagnosis of pigmented annular facial lesions**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Melasma</td>
<td>Photosensitive condition mainly seen on the face; common in pregnancy. Three typical distribution patterns: centrofacial, malar and mandibular</td>
</tr>
<tr>
<td>Discoid lupus erythematosus</td>
<td>Typical lesions are discrete, erythematous infiltrative plaques commonly seen on the face, neck and scalp. Lesions heal leaving depressed central scars, atrophy, pigment changes and telangiectasiae</td>
</tr>
<tr>
<td>Secondary syphilis</td>
<td>Dermatological clinical picture can take multiple forms: pustular, nodular, condyloma lata, maculopapular</td>
</tr>
<tr>
<td>Granuloma annulare</td>
<td>Self limiting, commonly with a rope-like border and central clearing; more common in females</td>
</tr>
<tr>
<td>Polymorphous light eruption</td>
<td>Pruritic rash occurring after sun exposure, resolving over days. More common in fair skinned people with an onset before the age of 30 years</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>More common in Afro-American people. Cutaneous involvement occurs in up to 20% and may present as nodules, plaques, maculopapular eruptions or thickening of old scars</td>
</tr>
<tr>
<td>Fixed drug eruption</td>
<td>Re-administration of drug leads to recurrence of plaques in same location; associated with hyperpigmentation</td>
</tr>
<tr>
<td>Erythema dyschoronic perstans</td>
<td>Asymptomatic, symmetrical, greyish pigmented irregular shaped plaques, most commonly seen in people of Latin American descent</td>
</tr>
<tr>
<td>Morphea</td>
<td>Two forms: localised (isolated sclerotic plaques) and generalised (symmetrical involving trunk and limbs)</td>
</tr>
<tr>
<td>Actinic lichen planus</td>
<td>Photosensitive condition, more common in people of Middle Eastern descent. Four recognisable morphological patterns (Table 2)</td>
</tr>
</tbody>
</table>

**Answer 2**

It is important to ask about risk factors and clinical features of syphilis including high risk sexual practices and the occurrence of a painless, self resolving, erosive and button-like papule (chancre), typically around the genital or oral regions. Discoid lupus erythematosus is usually limited to the skin, but can occasionally be found in systemic lupus erythematosus (SLE). Symptoms associated with SLE and sarcoidosis depend on the extent and type of organ involvement. Commonly reported symptoms in sarcoidosis include chest pain, dry cough, shortness of breath, dry mouth and eyes, as well as constitutional symptoms such as fever, fatigue and arthralgia. Commonly reported symptoms in SLE include fever, malaise, joint pain, and fatigue. A full medication history is vital.

**Answer 3**

It is important to examine the rest of the skin with a particular focus on sun exposed areas. If there is a history suggestive of syphilis, examine the genitals and mucosal surfaces for ulceration and look for lymphadenopathy. The most common clinical signs seen in patients presenting with sarcoidosis are neurological (14%), chest crackles (14%), and wheeze (9%).2 However, in patients with sarcoidosis, symptoms are more prevalent than signs at presentation. Systemic lupus erythematosus may affect many systems of the body and clinical findings could be multiple (including fever, arthritis, psychiatric disturbances, pericarditis, pleurisy and abdominal pain).

**Answer 4**

A skin punch biopsy specimen sent for histology and immunofluorescence is essential. The following tests may also be helpful in excluding a systemic cause of DLE, such as a manifestation of SLE and syphilis (sarcoidosis requires the finding of granulomas on biopsy from one or more sites):

- full blood examination (FBE), urea, creatinine and electrolytes (EUC), liver function tests (LFT)
- inflammatory markers (ESR, C-reactive protein [CRP]) and auto-immune markers including ANA, ENA and dsDNA
- syphilis serology

**Answer 5**

The most likely diagnosis is actinic lichen planus. The main differential diagnosis is DLE as both conditions may have similar clinical and histological presentations. The findings on immunofluorescence were nonspecific. Although positive linear deposits of immunoglobulins and C3 are suggestive of lupus, up to 20% of cases of DLE are negative, as in lichen planus.3 Similarly, inflammatory markers and autoimmune profiles are not relevant in differentiating the conditions. In this setting, the combination of clinical and histological findings assists in determining the final diagnosis.

In this case, the nonscarring characteristics of the facial lesions, negative autoimmune markers and immunofluorescence, as well as the presence of regular lichenoid features on histology without typically DLE features such as basement membrane thickening and follicular plugging, support the diagnosis of actinic lichen planus. In addition, 6 months later, Mrs AA developed well defined papules with shiny surfaces on the dorsum of her hands (Figure 2) as well as similar annular pigmented lesions on her arm. This supported the working diagnosis of actinic lichen planus.

**Answer 6**

Patients should be advised to use sunscreen and limit sun exposure. Treatment options include antimalarial agents, acitretin (an orally administered second generation retinoid) and corticosteroids. Consistently good results are noted with oral, intralesional and topical corticosteroids, however a mixed response is seen with antimalarials.4,5 Although up to 70% of patients could improve with acitretin, it is important to note that the teratogenic effect can...
last for up to 2 years. Biological agents such as topical pimecrolimus may also have a role in the management of this variant of lichen planus.5

Discussion
Actinic lichen planus is a variant of lichen planus that affects sun exposed areas of the body, most commonly the face, forehead, neck and extensor surfaces of the hands and forearm. Alternative names include lichen planus tropicus, lichen planus subtropicus, summertime actinic lichenoid eruption, and lichenoid melanodermatitis. It has a predilection for young adults of Middle Eastern descent with dark complexions. Actinic lichen planus represents about 15% of all cases of lichen planus in the Middle East.7 The condition is also more prevalent in people from African and Indian descent. In the Australian multicultural context it is important for clinicians to be aware of this variant of lichen planus to avoid misdiagnosing patients as having cutaneous lupus erythematosus.

The aetiology of actinic lichen planus is unknown, however ultraviolet radiation is thought to play a role. The use of repeated ultraviolet B irradiation has been reported to induce the lesions of actinic lichen planus.8 There are four morphological patterns (Table 2).9,10 Our patient presented with annular pigmented plaques and then subsequently developed classic lichenoid papules on the dorsal aspect of her hand and forearm (Figure 2). Actinic lichen planus can be distinguished from classic lichen planus based on clinical features (Table 3).8–10

Case follow up
Mrs AA’s lesions failed to improve with a course of hydroxychloroquine but completely remitted with a tapering dose of oral prednisone. Acitretin was not considered due to the teratogenic risk profile of the medication.

Summary of important points
- Actinic lichen planus is a variant of lichen planus that is mainly seen in young people of Middle Eastern, Indian and African descent.
- There are four described morphological patterns: annular, pigmented, dyschromic and classic lichenoid.
- Patient history and clinical examination assist in differentiating actinic from classic lichen planus and other differential diagnoses.
- Management includes advice on sun avoidance and sunscreen use and treatment with anti-malarial agents, corticosteroids or acitretin.

Authors
Chris Fessa MBBS, is Research Associate, Department of Dermatology, Westmead Hospital, New South Wales. cfessa@yahoo.com.au
Pablo Fernández-Peñas MD, PhD, FACD, is Associate Professor, University of Sydney, Sydney Medical School Westmead and Department of Dermatology, Westmead Hospital, New South Wales.

Conflict of interest: none declared.

Table 2. Morphological patterns of actinic lichen planus8–10

<table>
<thead>
<tr>
<th>Form</th>
<th>Clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Annular</td>
<td>Most common form: hyperpigmented, usually present over the face and dorsal aspect of hands</td>
</tr>
<tr>
<td>Pigmented</td>
<td>Melasma-like patches on face or neck</td>
</tr>
<tr>
<td>Dyschromic</td>
<td>Discrete or confluent whitish papules with a propensity to development into plaques, most commonly seen the posterior neck and dorsal aspect of hands</td>
</tr>
<tr>
<td>Classic lichenoid</td>
<td>Violaceous papules/patches, may develop in conjunction with other forms</td>
</tr>
</tbody>
</table>

Table 3. Difference in clinical features between classic and actinic lichen planus8–10

<table>
<thead>
<tr>
<th>Clinical feature</th>
<th>Classic</th>
<th>Actinic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Distribution</td>
<td>Usually involves flexural aspects of limbs, torso, mucosal membranes, nails</td>
<td>Photosensitive regions for example face and extensor surfaces of limbs, nil mucosal or nail involvement</td>
</tr>
<tr>
<td>Pruritus</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Köbner phenomenon</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Average age of onset</td>
<td>47 years</td>
<td>28 years</td>
</tr>
<tr>
<td>Seasonal variation</td>
<td>Nil</td>
<td>Increased incidence in Indian, Middle Eastern and African populations</td>
</tr>
<tr>
<td>Ethnic predilection</td>
<td>Nil</td>
<td></td>
</tr>
</tbody>
</table>

References

correspondence afp@racgp.org.au