Multiple facial plaques

A case study

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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 pigmentary 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: haemaglobin 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?

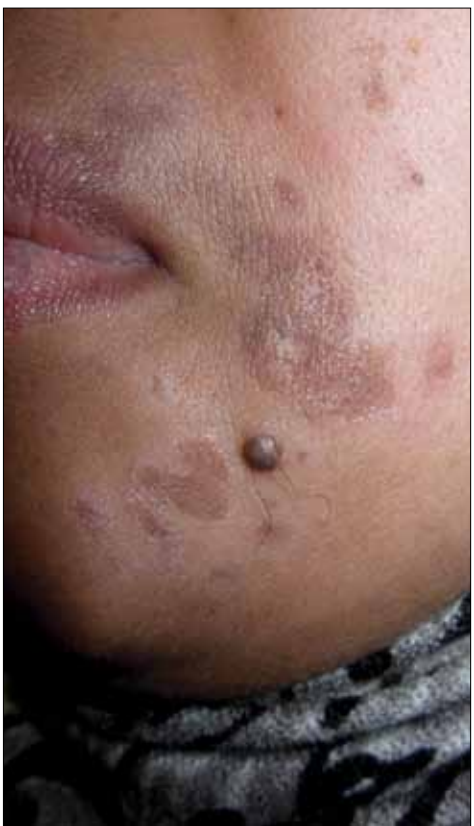

Figure 1. Small, nonpruritic, hyperpigmented annular plaques adjacent to the left side of the lip

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]

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 erythematosis 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).

Table 1. Differential diagnosis of pigmented annular facial lesions[1]

Condition	Features
Melasma	Photosensitive condition mainly seen on the face; common in pregnancy. Three typical distribution patterns: centrofacial, malar and mandibular
Discoid lupus erythematous	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
Secondary syphilis	Dermatological clinical picture can take multiple forms: pustular, nodular, condyloma lata, maculopapular
Granuloma annulare	Self limiting, commonly with a rope-like border and central clearing; more common in females
Polymorphous light eruption	Pruritic rash occurring after sun exposure, resolving over days. More common in fair skinned people with an onset before the age of 30 years
Sarcoidosis	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
Fixed drug eruption	Re-administration of drug leads to recurrence of plaques in same location; associated with hyperpigmentation
Erythema dyschronicum perstans	Asymptomatic, symmetrical, greyish pigmented irregular shaped plaques, most commonly seen in people of Latin American descent
Morphea	Two forms: localised (isolated sclerotic plaques) and generalised (symmetrical involving trunk and limbs)
Actinic lichen planus	Photosensitive condition, more common in people of Middle Eastern descent. Four recognisable morphological patterns (Table 2)

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

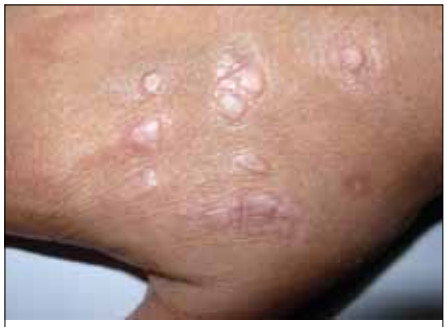

Figure 2. Multiple shiny, flat topped, firm papules resembling classic lichen planus on the dorsal aspect of the right hand

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.[6]

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.

Table 2. Morphological patterns of actinic lichen planus[9,10]

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

Table 3. Difference in clinical features between classic and actinic lichen planus[8–10]

Clinical feature	Classic	Actinic
Distribution	Usually involves flexural aspects of limbs, torso, mucosal membranes, nails	Photosensitive regions for example face and extensor surfaces of limbs, nil mucosal or nail involvement
Pruritus	Present	Absent
Köbner phenomenon	Present	Absent
Average age of onset	47 years	28 years
Seasonal variation	Nil	Usually appears in spring and summer and improves during winter
Ethnic predilection	Nil	Increased incidence in Indian, Middle Eastern and African populations

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.

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Conflict of interest: none declared.

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