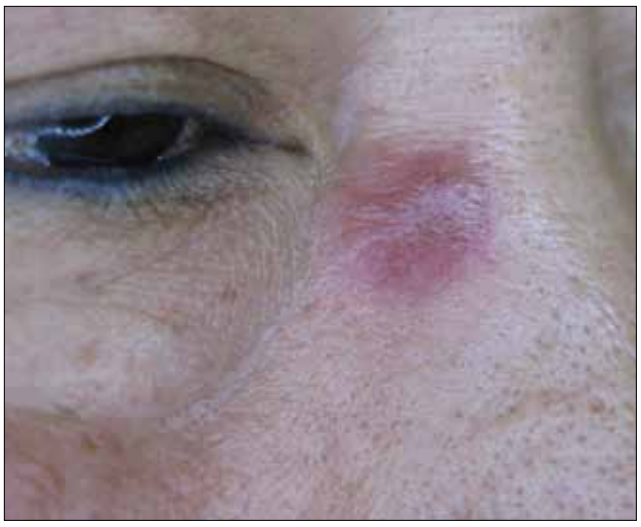
Answer to Dermacase *continued from* **page 977**

2. Sarcoidosis



Sarcoidosis is a chronic multisystem granulomatous disease that commonly involves the lungs. It affects people of all ages and races, with a slightly higher prevalence in women. Disease onset is typically between 25 and 35 years of age, with a smaller second peak in individuals older than 50 years. Diagnosis is often delayed because presentation is variable and symptoms are nonspecific; 30% to 60% of patients who have sarcoidosis with pulmonary involvement are asymptomatic.

Because the proliferation of lymphocytes and the recruitment of macrophages induce chronic granuloma formation, it is believed that there are 3 steps in the development of this disease: 1) An unknown antigen is presented by macrophages (or other antigen-presenting cells) to CD4 lymphocytes; 2) these activated macrophages produce interleukin (IL)–12, which is an important regulator of T-helper-1 immune response[1]; and 3) T-helper-1 lymphocytes secrete macrophage-stimulating cytokines, such as tumour necrosis factor–α, interferon-γ, and IL-2. Tumour necrosis factor–α and IL-2 recruit macrophages to the lesion sites, and interferon-γ transforms macrophages into giant cells.[2,3] However, the exact cause of sarcoidosis is still unknown.

Clinical presentation

Approximately 90% of sarcoidosis cases involve the lungs, but the skin, eyes, heart, joints, and lymph nodes are also commonly affected. At least 25% of patients with sarcoidosis exhibit cutaneous manifestations, which are either specific or nonspecific. Specific lesions often contain sarcoidal granulomas, while nonspecific lesions are reactive and without granulomas. Erythema nodosum is the most common nonspecific skin lesion associated with sarcoidosis. Interestingly, erythema nodosum, acute arthritis, and bilateral hilar lymphadenopathy comprise the typical triad of Löfgren syndrome, an acute and benign form of sarcoidosis.

Specific lesions frequently occur on the head and neck, and might present as papules, nodules, or indurated plaques.[4] Specific lesions are usually asymptomatic, but pruritus or pain can occur. One study has shown that sarcoidosis tends to affect women 40 years of age and older, and often manifests as chronic angiolupoid lesions, as was the case with our patient. **Table 1** outlines the typical clinical morphology of specific cutaneous lesions associated with sarcoidosis.[5]

Table 1. Specific cutaneous lesions associated with sarcoidosis

LESION	TYPICAL CLINICAL MORPHOLOGY
Papular	Translucent, firm, red-brown papules, mainly on the face and neck
Nodular	Red-purple nodules, frequently presenting on the extremities
Angiolupoid	Pink-red plaques with prominent telangiectasias, usually on the paranasal area; less numerous than those of lupus pernio
Lupus pernio	Relatively symmetric, red-purple shiny nodules and plaques on the nose, lips, cheeks, and ears

Diagnosis

Diagnosis of sarcoidosis is based on clinical manifestation, radiographic features, and histopathologic presentation of noncaseating granulomas. It is a diagnosis of exclusion: other granulomatous diseases, such as tuberculosis, atypical mycobacteriosis, fungal infection, foreign body reaction, or rheumatoid nodules, must first be ruled out. Family physicians should refer patients with persistent indurated facial lesions for histology and chest radiography. Physical examination, ophthalmologic consultation, electrocardiography, tissue biopsy, and laboratory tests for hepatic and renal function should be arranged for patients with sarcoidosis to check for possible systemic involvement.

Table 2 summarizes the disorders involving indurated facial lesions that should be differentiated from sarcoidosis.[5]

Treatment

Because sarcoidosis might spontaneously remit, and typical therapies carry the risk of adverse events, treatment of systemic sarcoidosis is only indicated when there is evidence of disabling symptoms or organ dysfunction. Corticosteroids are the first-line treatment; however, other immunomodulators, such as methotrexate, azathioprine, minocycline, hydroxychloroquine, thalidomide, cyclophosphamide, and infliximab, can be substituted in order to avoid steroid-induced side effects.[6] Cutaneous sarcoidosis can be treated with potent topical steroids, topical

Table 2. Clinical differential diagnosis of indurated facial plaques

DISEASE	TYPICAL CLINICAL MORPHOLOGY
Lupus vulgaris (cutaneous tuberculosis)	Red-brown plaques, with confluent papules
Discoid LE (chronic cutaneous LE)	Red, round plaques, with scaling and central atrophy
Tertiary syphilis	Dull-red, shiny, flat nodules and plaques, with crusts or ulcerations
Deep fungal infection	Red inflammatory plaques, with crusts or ulcerations

LE—lupus erythematosus.

tacrolimus, and intralesional triamcinolone injections. Oral medications should be reserved for large or disfiguring lesions. Skin biopsy should only be performed if there has been a poor response to topical treatment.

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Competing interests
None declared

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