

# Mauskopf facies

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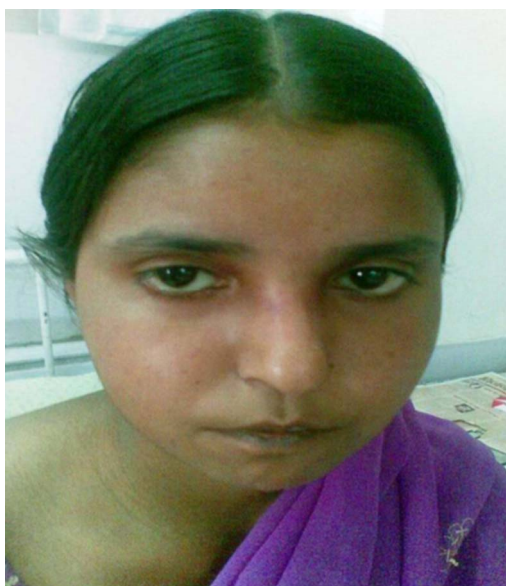
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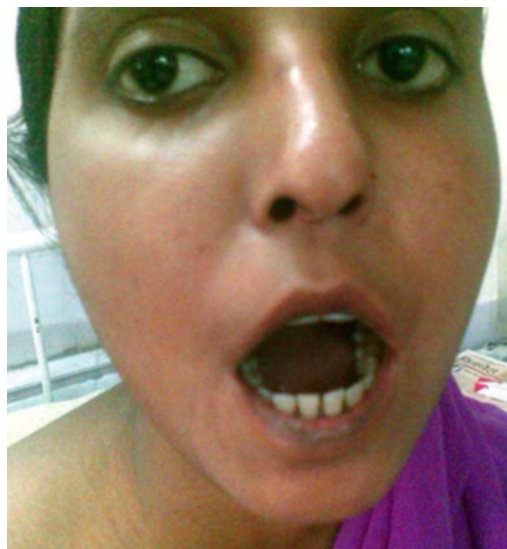
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## DESCRIPTION

An 18-year-old girl presented with a 15-month history of swelling in both hands and feet, decreased opening of mouth and ulceration of the tips of fingers. She also gave a history of bluish discoloration of the fingers and toes, and pain on exposure to cold. She had expressionless face (reduced mobility of eyelid, cheek and mouth) with taut, shiny skin and loss of wrinkles. Her nose was pinched with a beak like appearance (figure 1). There was thinning of lips (figure 1) with reduced oral aperture (figure 2). Fingers and toes were indurated and had ulceration and necrosis. Raynaud's phenomenon was demonstrated. A full thickness skin biopsy was suggestive of the expansion of the dermis due to the presence of thick bundles of collagen with loss of fat. Anticentromere antibodies were detected. The diagnosis of systemic sclerosis is usually made on clinical grounds in a patient with typical Mauskopf facies, Raynaud's phenomenon and digital changes.



**Figure 1** Expressionless face with taut, shiny skin and beak-shaped nose. There is also thinning of lips.



**Figure 2** Mouth opening was reduced.

A full thickness biopsy of the skin may be required in some patients. It is important to differentiate between the limited cutaneous and diffuse systemic type of subsets, as prognosis is better with the former.

## Learning points

- ▶ These constellations of skin changes, known as Mauskopf facies, are pathognomonic of systemic sclerosis.
- ▶ Their presences easily differentiate diffuse scleroderma from limited cutaneous variant.

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