



MORPHEA

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Scleroderma comes in two main forms: systemic and localized. **Scleroderma** is a disease of unknown origin that affects the microvasculature and loose connective tissues of the body and is characterized by fibrosis and obliteration of vessels in the skin, lungs, gut, kidneys and heart. Morphea is a localized form of scleroderma and affects primarily just the skin. Lesions are usually limited and most commonly just one lesion is found. However morphea can occur in a generalized form as well as guttate, nodular, subcutaneous and linear forms. Morphea is relatively uncommon and women are affected about three times as often as men. It is also more common in blacks and most patients are between the ages of 20 and 50 years of age at the time of diagnosis. Linear morphea is usually seen at an earlier age with most patients being under 20 years of age. Linear morphea can also affect not just the skin but the underlying structures such as muscle and bone. Antinuclear antibodies, elevated immunoglobulin levels and rheumatoid factor have been known to be elevated in all forms of localized scleroderma but the overall incidence is unknown. In spite of this the prognosis is generally good; the disease often becomes inactive in 3 to 5 years. Residual atrophy and hyperpigmentation/dyscoloration of the affected skin frequently remains. Disability from damage of underlying structure such as muscle atrophy can occur but is quite rare.

The appearance of morphea is that of a well defined ivory-colored plaque with varying degrees of inflammation about its borders. It generally starts as a reddened area that appears edematous. The center gradually becomes white or yellow in color. There can be a lack of hair noted in these lesions and sweating may be reduced depending on how advanced the lesion has become.

Generalized morphea is a severe form of the localized disease. Involvement of the skin is widespread with multiple plaques/lesions. The plaques may be larger than in the localized form but in the early stages they may be indistinguishable. It is not associated with systemic disease.

Guttate morphea may be a variant of **lichen sclerosus** and the two conditions can be seen in the same patient. It is characterized by small chalk white lesions that lack the firm center found in larger lesions of morphea. It involves primarily the neck and upper trunk areas of the body.

Nodular morphea (also known as keloid morphea) is quite rare. Nodular lesions are seen instead of plaques that resemble keloids. It can be seen along with more typical forms of morphea which aids in its diagnosis.

Subcutaneous morphea (also known as morphea profunda) is noted by deep, bound-down, sclerotic plaques. This occurs much deeper in the skin and because of this the typical inflammatory changes and coloration is not generally seen.

Atrophoderma of Pierini and Pasini is an uncommon very superficial form of morphea in which develops several oval, hyperpigmented (dark) atrophic plaques on the trunk. The distinctive feature is the lack of sclerosis seen in other forms of morphea. In a well developed lesion there is a slightly depressed center with a well defined border described as "cliff-drop" in appearance.

Linear morphea (linear Scleroderma) occurs in a linear band, usually only on one side of the body, not symmetrically. The most frequently involved areas are the lower extremities, followed by the upper extremities, frontal areas of the head and anterior torso. If this occurs on the face or forehead area it is known as 'coup de saber' or strike of the sword, and is characterized by atrophy with furrow formation of the skin and underlying structures. The atrophy can be quite marked with involvement of half the face, sometimes with atrophy of the tongue on the same side. The distinction between morphea and linear morphea is that morphea does not involve the underlying structures of the skin.

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The treatment of localized scleroderma remains unsatisfactory. Fortunately it is generally self-limiting and if it is an easily hidden area may require no treatment at all. The usual treatments involve the use of high potency **topical corticosteroids** or **intralesional corticosteroids**. Another treatment is the use of the vitamin D analog **calcipotriene** which is also applied topically. Other treatments could be considered but are best discussed with a dermatologist who is experienced in the treatment of scleroderma and morphea.

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