SYSTEMIC SCLEROSIS (SCLERODERMA) **MEDICAL APPENDIX**

DEFINITION

1. **Systemic sclerosis** is a multisystem disease in which the characteristic features are collagen proliferation, mild inflammatory-cell infiltration, obstruction of small blood vessels and ischaemic atrophy.

2. The term **'Systemic Sclerosis'** is preferable to **'Scleroderma'** as the latter suggests that the disease involves the skin alone. Indeed, in some cases, Systemic Sclerosis may not affect the skin. The term Scleroderma should be reserved for the purely cutaneous features.

CLINICAL FEATURES

3. The first features of the disorder usually appear between 30 and 50 years of age but may develop in any age group.

4. Raynaud's phenomenon (reversible peripheral ischaemia occurring predominantly in the hands but also in the feet and sometimes elsewhere, usually precipitated by cold) is by far the most common presentation and may be the only feature for many years.

5. Thereafter, there may be progressive induration and stiffening of the skin with loss of movement of the fingers.

6. Much less frequently, the presenting features may be polyarthritis, dysphagia, small bowel or other problems.

7. Skin changes vary from minimal changes in the fingers with minor tethering of the skin, to extensive involvement of the whole body, which may appear as if encased in a suit of armour. The term **'Sclerodactyly'** refers to changes limited to the fingers. **'Acrosclerosis'** describes the skin changes occurring predominantly in a peripheral distribution (hands, feet, distal forearms and legs. These may also include the face around the mouth). **'Diffuse Scleroderma'** implies involvement of the trunk.

AETIOLOGY

8. The disease may develop at any age, with a peak incidence in the fourth or fifth decade. It is rare in children. In young people there is relatively a much higher incidence in females than males (15:1), whereas in adults over the age of 45 the female to male ratio is only 2:1. It affects all races.

9. Although the aetiology of the condition is not known, certain factors are associated with its development –

    9.1 Exposure to various environmental factors may be associated with Systemic Sclerosis and an increase incidence in the condition has been reported in stone-masons, coal-miners and gold-miners.

    9.2 Systemic Sclerosis-like conditions have developed following exposure to vinyl chloride, other chlorinated hydrocarbons and epoxy resins.

9.3 Some drugs may produce Scleroderma-like conditions. In particular, Bleomycin, Tryptophan and Carbidopa have been identified. Features of Scleroderma have been reported after paraffin wax implantation for cosmetic surgery.

9.4 There has been a number of familial cases of Systemic Sclerosis and an increased incidence of some tissue types has been associated with some of the varieties of the disease.

9.5 Associations have also been reported with Diabetes Mellitus, particularly the insulin dependant, childhood variety.

9.6 Auto-immunity may play some part in the development of the disease and, in some cases, there may be a genetic pre-disposition.

CONCLUSION

10. **Systemic sclerosis** is a multisystem disease in which the characteristic features are collagen proliferation, mild inflammatory-cell infiltration, obstruction of small blood vessels and ischaemic atrophy. Whilst its aetiology is unknown, certain factors which have been listed above are associated with its development.

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