EDITORIAL

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SCLERODERMA

The term scleroderma has been derived from the Greek language and it means hardening of the skin. Numerous accounts of scleroderma appeared in the nineteenth century but it may have occured earlier than that. Towards the middle of the twentieth century it became appreciated that affections of skin and vessels are nothing but manifestations of a general disease which may involve many organs.

Although not very common, this disease has wide racial and geographical distribution. In the tropical countries, lack of recognition may be the real factor responsible for its apparent dearth.

The exact aetiology is not known. The disease does not run in families. Middle aged women are more commonly affected than others. Persons exposed to silica dust are prone to develop this disease but this is not true for the majority of patients. No infective agent can be held responsible for the disease although the early pathological changes are inflammatory in nature. The main brunt of the attack is borne by the skin and blood vessels although it is more appropriately known as a multisystem disease with predilection for particular organs, perivascular cellular infiltration exclusively of a mononuclear character and hyperaemia are the early findings in a skin lesion. Collagen bundles are swollen, homogenous and separated by odema and cellular infiltrate of lymphocytes. Most of the other pathological features can be explained by the intimal thickening and reduction in the lumen of blood vessels leading to parenchymatous degeneration or replacement by fibrous tissue. Accumulation of inflammatory cells and occurrence of degenerative changes may be found in skeletal muscles as well as cardiac muscle. The changes in the kidney are most striking because the pathological changes are similar to those in malignant hypertension without the concomitant rise in blood pressure. The chief constituent of connective tissue is collagen and therefore electron microscopic studies of this substance have been carried out but not much abnormality of collagen fibrils have been demonstrated. Hence, further research was aimed at the other component of connective tissue, namely ground substance. It has been suggested that disturbance of mucopolysaccharides may be of primary importance. Abnormal catecholamine metabolism and serotonin excess have also been postulated as probable causes. A significant finding is that collagen like plasma protein called hypro-protein is in excess and urinary hydroxyproline output is also more in patients suffering from this disease particularly if duration of the disease is for less than a year.

There are suggestive evidences that scleroderma may be an auto-immune

disease but the difficulty is that virtually all our knowledge of immune tissue injury is confined to cells as the injured party[1]. In this case, it is possible that collagen itself or its associated ground substance provides the antigenic stimulus. Generally accepted classification of scleroderma[2] is (1) Systemic scleroderma i.e. progressive systemic sclerosis and (2) Localised scleroderma which is further subdivided into morphoea and linear scleroderma. Besides, there are sclerodermoid lesions. The advantage of classifying the disease into Type-1, 2 and 3 depending on the extent of the disease is that prognostically type 1 disease has a normal life expectancy and type 3 disease usually proves fatal within five years.

In addition to skin sclerosis, calcinosis and telangiectasia may be found in some cases.

Reference to multisystem involvement has already been made. Joint involvement or visceral involvement predominates in some cases along with skin involvement. Oesophagus, intestines, heart, lungs and kidneys are the most frequently involved viscera.

Oesophageal and intestinal involvement generally give rise to obstructive features. The peculiarity of lung involvement is that although there may not be any clinical or radiological findings, functional impairment may be so much that the patient may be a respiratory cripple or even succumb to the disease.

Cardiac abnormalities in scleroderma are either due to replacement of cardiac muscle by connective tissue or secondary to increased pulmonary vascular resistance. Pericarditis, arrhythmias and even cardiac failure may be precipitated. Renal changes have already been referred to. Mild chronic impairment of renal function due to scleroderma has not been extensively studied but acute unheralded renal failure associated with hypertension has, for obvious reason, allowed detailed study of the characteristic pathological picture in the kidneys. Central Nervous System disturbance is rare and probably occurs secondary to involvement of blood vessels supplying the brain.

Diagnosis of scleroderma is mainly clinical. Radiological investigations are, of course, carried out in cases of visceral involvement. For confirmation of the diagnosis histological examination of biopsy material is carried out. It would be of interest to quantitate the degree of skin thickening by taking circular biopsies of uniform surface diameter (7 mm) from the dorsal aspect of the forearm using a dermal punch. Prof. Gerold P. Rodnan[3] advocated this method at the W. H. O. Conference on scleroderma held in Paris in 1969. Apart from the absolute and proportionate increase in the quantity of dermal collagen, qualitative abnormality in the shape of slight increase in collagen-bound hexoseamine is found in such cases. The biopsy material may further show the presence of antinuclear factor of the speckled variety.

Despite all these, it must be admitted that diagnosis may sometimes be delayed by two years in type-1 disease when the sclerosis is limited to fingers. In the absence of knowledge of the cause and pathogenesis of the disease, there is no curative treatment. Symptomatic treatment is the mainstay although steroids are extensively used at least for temporary regression of skin changes. It must, however, be appreciated that steroids can not influence the visceral disturbances or the longevity of the patient.

In recent times, penicillamine and progesterone-like drugs have raised our expectation in view of their known effect on collagen metabolism. There has been a report of successful treatment by haemodialysis, nephrectomy

and renal transplantation in a case of scleroderma with renal failure. This is an isolated instance and it does not provide a clue to the treatment of the condition known as progressive systemic sclerosis. Until the cause is known, relentless efforts are necessary to find out more about this disease by research workers particularly in the fields of histochemistry and immunology.

References

1. Glynn LE: Scleroderma, Proceed Int Symp sponsored by W H O, Paris-VI, 1972, p 280

2. Tuffannelli DL, Roed WB and Stepani J : Scleroderma, Proceed Int Symp sponsored by W H O, Paris-VI, 1972, p 16.

3. Gerald RP: Scleroderma, Proceed Int Symp sponsored by W H O, Paris-VI, 1972, p 271.

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TRUE or FALSE ?

Epidermodysplasia verruciformis (EV) is induced by human papilloma/virus of which there are 2 distinct types - HPV - 3 and HPV - 4

(Answer Page No. 264)