

## 22. Scleroderma

### LEARNING OBJECTIVES:

1. Understand the multifaceted pathobiology and pathophysiology of scleroderma as it relates to endothelial dysfunction and the selective expression of various cytokines resulting in distinct alterations in function of various cell lineages.
2. Recognize the unique clinical, laboratory, and radiologic features that contribute toward the establishment of the diagnosis of scleroderma.
3. Understand that the therapeutic approaches to treating scleroderma relate specifically to the organ system involved and to the severity of physiologic dysfunction.

### SUMMARY:

1. Scleroderma is diagnostically subdivided into “limited” and “diffuse” subtypes defined according to the extent of cutaneous involvement.
2. Both limited and diffuse scleroderma have unique internal organ involvement which is relatively specific to each subtype.
3. Although the end result of the sclerodermatous process is fibrosis of target organs, the primacy of vasculopathic mechanisms in this process is highlighted by the invariable presence of Raynaud’s disease at the onset of disease.
4. Aggressive immunosuppression has a very limited role in treating the clinical manifestations of scleroderma since, for the most part, this has not been shown to have had a significant effect on the morbidity or mortality of the disease.