



# Emergency Management of Scleroderma, Raynaud's and Lupus

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# Emergency Care

- Three things to remember in the Emergency Department
- Don't assume you know more than your patient
- Listen to the patient
- It's not about you, it's about the patient

# Scleroderma

- Scleroderma is characterized by both a limited and diffuse variant
- Characteristic skin thickening with visceral involvement heralds the diffuse variant
- For Emergency purposes it is visceral involvement complications that have important emergent consequences

# Scleroderma

- From an emergency perspective it is important to be reminded of a few key components of care as it relates to scleroderma
- Also important that in the case of rare diseases that often the patients and family may have a better understanding of the disease than the provider
- Ask for help when indicated

# Raynaud's

## Phenomenon

- Commonly seen in scleroderma
- Can present with digital ulceration and gangrene
- Treatment includes Calcium Channel Blockers, ACEII inhibitors, prostacyclin analogues, phosphodiesterase inhibitors and surgical and chemical sympathectomy
- Also amputation is sometimes indicated and treat infection aggressively
- Keep hands warm, avoid sudden changes in temperature

# SkinFibrosis

- Immune suppressing drugs such as methotrexate, cyclosporine, tacrolimus are often used
- Important to remember that any patient with a fever who presents to an Emergency Department who is immune suppressed is septic until proven otherwise
- Those who are immune suppressed often appear clinically well until late sepsis

# Arthritis and Myositis

- Can use NSAIDs, low dose steroids or methotrexate
- In Myositis immune suppression is used

# GI Involvement

- Esophageal motility most frequently encountered
- Mechanical advice, PPI's, pro kinetic agents and calcium channel blockers used
- Again important to consider other illness, rarely effects the biliary tree



# Renal Crisis

- Up to 10% of patients will experience malignant hypertension
- Cornerstone of treatment is with ACE inhibitors
- Key is to treat aggressively and prevent renal injury

# Pulmonary

- Both may be seen in scleroderma and are difficult to treat
- Hypertension and ILD
- In ER may present as acute SOB or L and R sided heart failure
- Also pericarditis can be seen and standard treatment is recommended
- Arrhythmia are also noted as well as myocarditis
- Left sided heart failure due to fibrosis in universally fatal

Remember! Multi  
system disease and  
immune suppression!

# Lupus

- Autoimmune disease
- Multi system disorder
- More common in women of child bearing age
- 1:600 white women, 1:200 black women
- 8 to 13 times more common in women

# Lupus

- Again multi-system disease often treated with immune suppression
- Any immune suppressed patient with Lupus and a fever is septic until proven otherwise
- May present with mimics in different organ systems

# Neuropsychiatric Lupus

- CNS Involvement is common in Lupus
- Patients particularly those with anti phospholipid antibodies can present with stroke like symptoms
- Patients can present with acute multi-level neuritis
- Also headaches are common

# Neuropsychiatric

- Also may present with depression, cognitive impairment, seizures, and psychosis
- Consider other causes but remain open to the diagnosis of autoimmune phenomena as cause for unexpected psychiatric symptoms

# Renal Lupus

- Lupus nephritis is common but if properly treated and recognized ESRD occurs in only 5% of cases
- Presents with hematuria, proteinuria, and often has a characteristic membranous GN
- Can be treated with steroids and cyclophosphamide



# Cardiac

- SLE can present with myocarditis, pericarditis and non infective endocarditis, particularly of the mitral and tricuspid valves
- Standard treatment is indicated but recognition and awareness are important

# Lupus

- Other common areas include the skin (classic malar rash) and joints
- Doesn't tend to cause joint deformities and tends to affect the small joints of the hands and feet

# Summary

- Be aware of multi system involvement in these conditions
- Be careful when patients are immune suppressed
- Ask for help