

VIDEO CASE SOLUTION

Solution: Nephrogenic Systemic Sclerosis (NSS)

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A 50-year-old woman with chronic renal failure and has been on hemodialysis for five years. She had multiple gadolinium enhanced MRIs for chronic low back pain (LBP). She developed inability to raise arms and legs that evolved over six months. She had no dysphagia. She was found to have hard skin and breasts and her CK level and EMG were normal.

The most likely diagnosis is:

1. Scleroderma
2. Polymyositis
3. Nephrogenic systemic sclerosis (NSS)
4. Polymyalgia rheumatica
5. Systemic calcinosis

- NSS is a dramatic hardening of the skin and subcutaneous tissue that is reported in uremic patients who get gadolinium contrast. The deposited mucin, collagen and elastic fibers are produced by activated fibroblasts.
- There is evidence that sclerosis goes beyond the skin and subcutaneous tissue. Muscle involvement is documented. Reported axonal neuropathy may also be related to uremia.
- CK level is usually low due to decreased muscle mass and EMG is either normal or shows mixed short and long duration units consistent with chronic myopathy.
- Other disorders with similar picture include amyloidosis, scleromyxedema, systemic sclerosis, eosinophilic fasciitis, and graft vs. host disease should be considered.
- The exact pathogenesis is not clear and some patients with uremia develop NSS without being exposed to gadolinium.
- Renal transplantation may improve outcome, otherwise, there is no cure for this condition. Steroids, PLEX, and IVIG are reported to cause transient improvement. ■

Neuromuscular involvement in nephrogenic systemic fibrosis. Keyrouz S, Rudnicki SAJ Clin Neuromuscul Dis. 2007 Dec;9(2):