

CLINICAL IMPROVEMENT IN NEPHROGENIC SYSTEMIC FIBROSIS (NSF) FOLLOWING RENAL TRANSPLANTATION: SHOULD 'NSF POINTS' BE CONSIDERED IN ALLOCATION?

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NSF is a fibrosing disease with serious consequences including limb contractures, multi organ involvement and high mortality. Gadolinium exposure is implicated. Treatment has been largely disappointing.

A 62 year old female on peritoneal dialysis for 18 months due to ESRD secondary to hypertension and chronic NSAID use underwent an MRI with contrast for work up of a complex renal cyst. Four weeks later, the skin over her lower extremities became sensitive to touch with burning sensation and periodic sharp shooting pains, with progressive tightening of her skin with time. On exam, the skin appeared tight and hyperpigmented with linear bands extending up to the lower thigh. Biopsy showed thickened and haphazardly arranged reticular dermal collagen bundles with prominent clefts, consistent with NSF. The skin tightness progressed to involve both her lower extremities entirely. She received a cadaveric renal transplant in July 2007. In the several weeks post transplant, the patient has reported improvement in the swelling of her lower extremities, and that she can actually pinch the skin over her thighs, and this was also confirmed clinically using a pinching score. A repeat skin biopsy is planned.

NSF is characterized by thickening and hardening of the skin and can also involve other organs. Skin lesions are variable, and can lead to joint contractures. Apart from renal failure, Gadolinium exposure during MRI/MRA in patients is the most consistent risk factor. Free Gadolinium is liberated from its chelates upon administration to patients with CKD, and is highly toxic. It is thought to undergo transmetallation in vivo, with other metals such as Copper, Zinc and Calcium, and once phagocytosed by macrophages, is associated with an increase in profibrotic cytokines or growth factors leading to dermal or systemic fibrosis.

There is no proven effective cure for NSF with only a return of renal function documented to be effective. Since this condition is relentlessly progressive, thought may be given for these patients to receive extra points in the renal allocation schema to receive timely transplantation.