Nephrogenic Systemic Fibrosis, also known as nephrogenic fibrosing dermopathy, is a recently recognized rare disease usually occurring in patients with renal insufficiency who were exposed to gadolinium containing contrast agents, and until recently, considered limited to the skin. The characteristic cutaneous findings include symmetrical skin thickenings on the extremities with accumulation of collagen and fibrous tissue, and often positivity for CD34, CD68, and Factor XIIIa. Although there have been recent reports of systemic involvement, bone marrow findings have not been reported to our knowledge.

Diffuse reticulin fibrosis was found in the bone marrow of this patient with nephrogenic systemic fibrosis, with an associated peripheral blood pancytopenia. Other etiologies for bone marrow reticulin fibrosis (such as myeloproliferative disorders, hairy cell leukemia, and human immunodeficiency virus) were excluded. The immunostaining pattern was not completely consistent with some reported cases of nephrogenic systemic fibrosis. This lack of immunostain similarity could either be due to site specific variations in disease presentation, or due to his extended disease course and therapy. The possibility of bone marrow involvement by nephrogenic systemic fibrosis leading to pancytopenia holds important diagnostic and treatment implications.

References