Nephrogenic Fibrosing Dermopathy; A Case Report

Reza M Robati1*, Somayeh Hejazi1, Zahra Asadi-kani1

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Abstract

Nephrogenic fibrosing dermopathy or nephrogenic systemic fibrosis (NSF) was first recognized in 1997, is a rare, diffuse, multisystem, fibrosing disease with an incidence of approximately 3–5% which affected men and women equally and its etiology remains to be defined. We describe a 45-year-old Iranian woman on renal transplantation who presented with multiple hypo and hyperpigmented, edematous, indurated, thickened plaques of her upper and lower extremities started 3 months after kidney transplantation with diagnosis of NSF.

Keywords: Nephrogenic fibrosing dermopathy; Nephrogenic systemic fibrosis; Renal failure; MRI

1. Introduction

Nephrogenic fibrosing dermopathy or nephrogenic systemic fibrosis (NSF) is a rare, diffuse, multisystem, fibrosing disease develops in the setting of advanced renal failure and characterized with a wide ranges of manifestations, from only mild skin manifestation to severely debilitating fibrosis that involving skin and other organs including the subcutaneous tissues, muscles, lungs, pleura, pericardium, and bones.(Shabana et al., 2008;Robinson et al., 2011; Kribben et al.,2009). In our patient, skin symptoms were relatively minor and were tolerated by the patient for as long periods as 7 years without definitive evaluation.

2. Case report

A 45-year-old woman presented to our outpatient clinic with a complaint of multiple plaques on her extremities which had been present since 7 years ago. Skin changes started on her left leg 3 months after renal transplantation, then spread to both legs, dorsal of foots, arms and dorsal aspect of the hands with a slowly expanding course. The plaques were non-tender and non-pruritic.

*Corresponding e-mail: rezarobati@sbmu.ac.ir
1 Skin Research Center, Shahid Beheshti University of Medical Sciences, Shohadae-Tajrish Hospital, Tehran, Iran
Physical examination revealed multiple symmetrical large hypo and hyperpigmented edematous and mildly indurated plaques on her forearms, dorsal aspect of the hands, legs, ankles and dorsal of feet with a pale and peau d’orange appearance [Figures 1 and 2].
Fig 2. Large indurated plaques on the forearm with a pale and peau d’orange appearance (closer view).

The joint examination showed normal range of motion, without sign of arthritis or muscle contraction. The mucous membranes and the other areas of the skin were intact. She had no evidence of systemic involvement or coexisting diseases, except positive history of hypertension and diabetes mellitus. A history of administration of gadolinium-based contrast for imaging was unremarkable for our patient.

Her family history was unknown. During these years, she has had neither evaluation nor treatments for these lesions. Two skin biopsies were performed from her leg and her arm. [Figure 3] The differential diagnosis of myxedema, mucinosis and generalized granuloma annularis was considered. Histopathological examinations revealed dense dermal fibrosis and sclerosis involved dermis, septa of subcutis and fascia, increased dermal spindled cells with scattered multinucleated giant cells compatible with clinical diagnosis of nephrogenic fibrosing dermopathy.
3. Discussion

Nephrogenic fibrosing dermopathy characterized clinically by thickening and hardening of the skin of the extremities and trunk with large brownish hyperpigmented edematous, fibrotic or woody plaques with or without pruritus and pain which may be mistaken for other fibrosing skin disorders e.g. scleroderma, scleromyxoedema, systemic sclerosis/morphea, pretibial myxedema, eosinophilic fasciitis and eosinophilia myalgia syndrome. (Kribben et al., 2009; Foss et al., 2009; Mendichovszky et al., 2008)

At risk patients included who had a renal failure, a history of previous exposure to gadolinium-based contrast material for MRI, especially in persons with higher dose of contrast exposure, elevated plasma level of iron, calcium and phosphate, presence of chronic metabolic acidosis, immunosuppression and vasculopathy, hypercoagulability, and tissue injury due to surgical procedures. (Shabana et al., 2008; Perez-Rodriguez et al., 2009) Unfortunately, a history of administration of gadolinium-based contrast before symptom onset was unknown for our patient, but in whom nephrogenic systemic fibrosis developed 3 months after renal transplantation.

The diagnosis of NSF may be confirmed through the histopathological examination that may show proliferation of fibroblast-like dermal cells, collagen remodeling, thickening and separation of collagen bundles by clefts and variable amounts of mucin deposition. (Kribben et al., 2009; Foss et al., 2009; Mendichovszky et al., 2008; Song et al., 2009)
Although unusual, spontaneously resolution of skin lesions was reported especially in persons with acute and transient renal failure. (Shabana et al., 2008)

According to the current knowledge, restoration of renal function by renal transplantation or recovery from acute renal failure is the main therapeutic goal, but not effective in all patient. (Kribben, et al., 2009) Other various therapeutic options that have been tried in the treatment of NFS with different success rate included extracorporeal photopheresis, systemic corticosteroids, plasmapheresis, ultraviolet phototherapy, intravenous immunoglobulin, thalidomide, pentoxyphyllin, cyclophosphamide and the chelating agent. (Kribben et al., 2009; Mendichovszky et al., 2008)

The findings in our case were similar to the findings of earlier experience with nephrogenic systemic fibrosis that correlated with compromised renal function, but the strongly correlation between gadolinium-based contrast administration and development of condition in this case was difficult to establish.

Because of the risks of gadolinium-based contrast material in renal failure patients, MRI contrast agents should be administrated with caution in these patients. It should be remembered that diagnosis of NFS must be considered in cases of enlarging plaques in the setting of renal disorders.

References


