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Nephrogenic fibrosing dermopathy – a rare case

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Abstract

The aim of the article is to present a rare case of nephrogenic fibrosing dermopathy (NFD) in a 54 year old male, which has been recently described fibrosing disorder that emerges in different settings of renal insufficiency. These are characterized by skin- coloured to erythematous papules that coalesce into plaques markedly thickened in texture. To date, the registry for NFD lists approximately 175 cases worldwide. In our case, clinical features, histopathology, special stains and immunohistochemistry were diagnostic of NFD.

Key Words: Nephrogenic Fibrosing Dermopathy, Chronic Renal Failure, Histopathology.

INTRODUCTION

Nephrogenic fibrosing dermopathy (NFD) was first reported in 2000 by Cowper's et al.[1] NFD is a recently described fibrosing disorder that emerges in different settings of renal insufficiency. The degree and cause of the underlying renal impairment seem not to be related to the severity of the nephrogenic fibrosing dermopathy.[2]

NFD characteristically presents symmetrically on the extremities and trunk. A common distribution is between the ankles and mid-thighs and between the wrists and mid-upper arms. These are characterized by skin- coloured to erythematous papules that coalesce into plaques markedly thickened in texture. To date, the registry for NFD lists approximately 175 cases worldwide.[3]

CASE PRESENTATION

A 54 year old male on hemodialysis for chronic renal failure, presented with large brownish indurated plaques on both the lower limbs. On examination, oedema & focal hyperpigmented patches were noted over lower limb (Figure-1).

Laboratory findings -: The urea nitrogen was 40 mg/dL (range 7-30), creatinine 5.5 mg/dL (range 0.5-1.2), and normal white-cell count 4.5 x 10^3/mcL. A lipid profile and liver function tests were within normal limits. HIV and HBs Ag, and Anti nuclear antibodies were negative. Skin biopsy was sent for histopathological evaluation.

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HISTOPATHOLOGY

Light microscopy

Epidermis showed hyperkeratosis, irregular acanthosis & follicular plugging. Dermis showed diffuse fibrosis (Figure-2). Also noted focal areas of dermal stromal mucin and intercalating spindle-like cells with sparse lymphocytic infiltrate and no adnexal structures were noted (Figure-3).

PAS stain

Showed focal positivity for mucin , differentiating it from more abundant mucin seen in scleromyxoedema (Figure-4).

Orcein stain

Showed increased numbers of elastic fibers.

Immunohistochemistry

Showed prominent CD68 positivity suggesting that the plaques were more than 20-weeks old (Figure-5).

DISCUSSION

Nephrogenic fibrosing dermopathy (NFD) is a rare condition in patients with renal insufficiency, which is characterized by diffuse hardening and thickening of the skin. Our case also reveals same finding of renal insufficiency & hardening of skin.

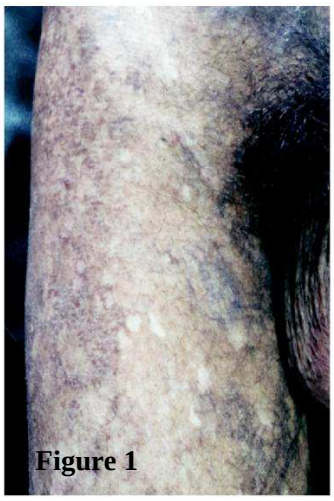
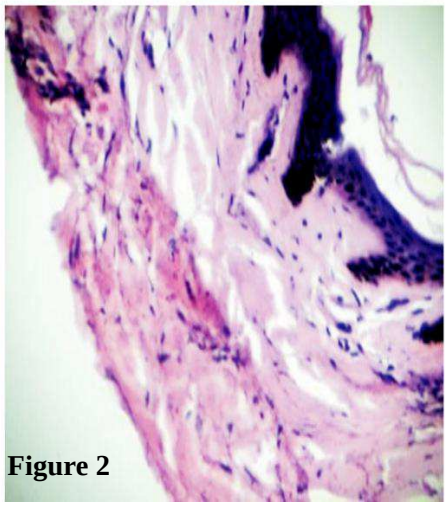


Figure 1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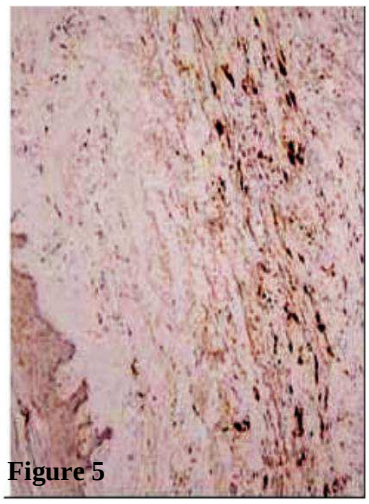
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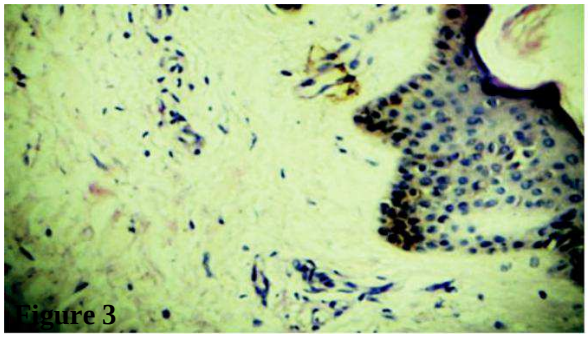
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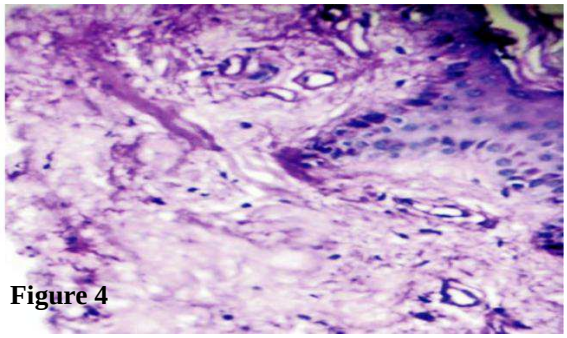
Figure 4

Figure 1 Clinical photograph showing, light brownish indurated plaque and edema over lower limb.Figure 2 Photomicrograph shows diffuse fibrosis. (H&E, 10X).Figure 3 Photomicrograph shows intercalating spindle cells & no adnexae. (H&E, 10X).Figure 4 Photomicrograph showing focal PAS positivity. (PAS,10X).Figure 5 Immunohistochemistry showing CD68 positivity.

The etiology and pathophysiology of NFD is unknown. The recent emergence and clustering of cases at major medical and renal transplant centers suggests a toxic or infectious agent, although no studies confirm.

A recent article raises the question of whether gadolinium-enhanced imaging studies, which are typically performed 2-4 weeks prior to disease onset, may trigger the development of NFD in susceptible persons [4].

Approximately 15 percent of patients have had an associated antecedent surgical procedure, most often a vascular reconstructive procedure. An appreciable number have chronic liver disease and some have idiopathic pulmonary fibrosis.[5]

The clinically and histologically NFD should be differentiated from scleromyxoedema, scleroderma & eosinophilic fasciitis.[6] Scleromyxoedema affects face (spared in NFD) and associated with systemic involvement & paraproteinemia. In our case face was spared and lower limbs were affected and clinical features, histopathology, special stains and immunohistochemistry were diagnostic of NFD.

The patient is under follow up and is keeping fine without any contractures.

CONCLUSION

NFD in patients with chronic renal failure of unknown cause is a poor prognostic indicator. So, early detection before the development of contracture and prompt treatment of NFD and underlying renal failure may reverse this disabling condition.

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