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**Case Report**

# Renal cyst occurring in a case of Darier's disease

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## ABSTRACT

Darier's disease is a rare genodermatosis inherited in an autosomal dominant pattern. The disease is characterised by chronic persistent hyperkeratotic papular eruptions over the seborrheic regions.

**Keywords:** Dariers disease, Genodermatosis, Renal cyst

## INTRODUCTION

Darier's disease or Darier- White disease, also known as Keratosis follicularis; was first reported in 1889 by Darier and White independently.[1] Renal involvements in the form of renal agenesis, polycystic kidneys and horseshoe kidneys have been reported. Here we report a case of Darier's disease with a renal cyst.

## CASE REPORT

We report a 52 year old male who came to the Dermatology OPD with diffuse pigmentation along with raised skin lesions for the past 6 months. The lesions first appeared over the axilla and then spread to involve the neck, back, scalp, forearms and legs. Similar lesions later developed over the face. The patient also complained of itching and photosensitivity. There was no history of similar lesions in the past, or in the family.

Local examination showed multiple warty papules on the scalp, forehead and nape of neck. Diffuse pigmentation of the face, neck and extremities were noted. Toe nails showed longitudinal ridging and nicking. Oral cavity was normal.

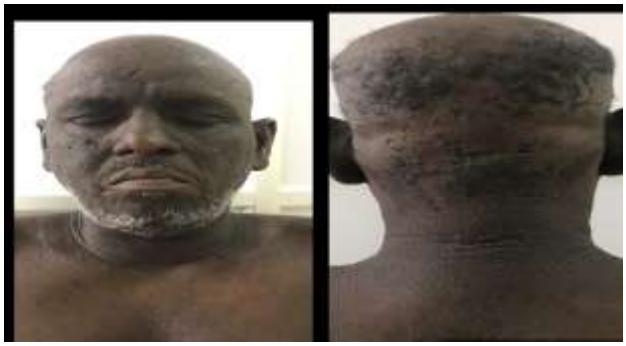

**Figure 1: Clinical picture showing warty pigmented papules on the face, nape of neck and scalp.**

An initial diagnosis of photo dermatitis was made while keeping Darier's disease into consideration. Skin biopsy showed hyperkeratosis with follicular plugging, few vacuolated keratinocytes, focal basal cell degeneration and increased pigmentation in the upper dermis. The epidermis also showed areas of suprabasal separation with corp ronds and grains. The histopathological picture was consistent with Darier's disease. Ultra sonogram of the abdomen revealed a cyst measuring 2.9 × 2.2 cm in the lower pole of the left kidney.

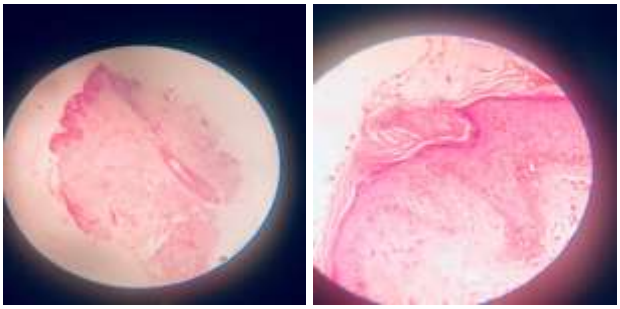

**Figure 2: Histopathology shows suprabasal detachment of epidermis at one end and hyperkeratosis along with follicular plugging on scanning view and follicular plugging along with corp ronds and grains on high power.**

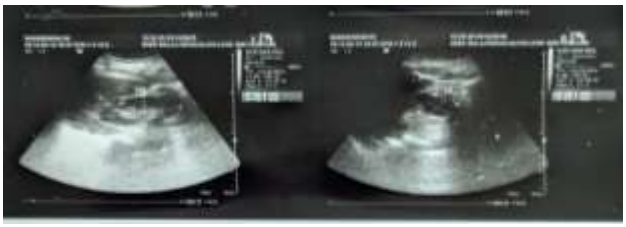

**Figure 3: Ultra sonogram showing left renal cyst.**

## DISCUSSION

Darier's disease is characterized by brown red keratotic papules which coalesce to form verrucous plaques.[2] The main Pathophysiology lies in the mutation of the ATP2A2 gene that encodes calcium transport pump, sarco/endoplasmic reticulum $Ca^{2+}$ adenosine triphosphatase pump.[3] SERCA2 is responsible for the transport of calcium ions from the cytosol into the endoplasmic reticulum, thereby maintaining a low cytoplasmic $Ca^{2+}$ levels.

Darier's disease affects both sexes equally. The rash appears during the teenage years mostly but there are times when they manifest for the first time during adulthood also.[4] The cutaneous lesions also include distinctive nail features like longitudinal ridging with V-shaped nicking and Palmoplantar abnormalities.[5]

Histopathological picture shows supra basal lacunae. Epidermis shows hyperkeratosis, papillomatosis, rounded dyskeratotic cells with eosinophillic cytoplasm called corp ronds and small cells with shrunken cytoplasm called grains[6].

The main differential diagnosis includes Seborrheic dermatitis, Hailey- Hailey disease, and Dowling Degos disease and Acanthosis Nigricans.[7]

Treatment options are based on the severity of the disease manifestations. Emollients, sunscreens, and light clothing are all that may be required for milder forms of the disease. The more severe forms call for the need of systemic steroids and oral retinoids.[8] Oral antibiotics are required in cases of secondary infection. Oral vitamin supplements like vitamin A and E have showed promising results.

## CONCLUSION

Darier's disease is a rare disorder of keratinisation with rare associations of renal disease. This is the second case being reported by one of the authors regarding the association of Darier's disease with renal cysts.[9] With the increasing number of such cases being reported in literature it can be concluded that this association is not merely coincidental; advocating the need for screening the renal system in patients with Darier's disease by imaging to rule out this possibility.

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