CASE SERIES

NEKAM'S DISEASE: THREE CASES FROM A FAMILY WITH AUTOSOMAL RECESSIVE INHERITANCE

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ABSTRACT
Nekam's disease, also known as Keratosis Lichenoides Chronica is an uncommon, keratinizing disease of skin and mucosa. Flat-topped violaceous papules & nodules on the dorsum of the extremities are its defining features. Reticulate, linear lesions along with telangiectasias and scaling involves seborrheic areas. Herein, we report three cases from the same family, with Nekam's disease. There have been a few case reports published, making it a rare disease.
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INTRODUCTION

Nekam's disease or Keratosis Lichenoides Chronica (KLC) is an uncommon, mucocutaneous condition of unknown etiology[1]. It appears in 20-50 years of age and affects both male and females. Clinically it presents with violaceous papules and nodules arranged in reticulate and linear pattern on the dorsal aspects of hands, feet, extremities and buttocks. Seborrhea like scaly rash with visible telangiectasia over the scalp, neck and face may be present[2]. It follows a chronic, progressive course[3]. It was initially called as "Lichen Planus and Lichen Ruber Acuminatus Morbilliform disease" by Kaposi, in 189 Nekam[4] named it "Porokeratosis Striate Lichen" on observing palmoplantar hyperkeratosis in the case reported by Kaposi[5]. In 1972 Margolis et al[3] introduced the term Keratosis Lichenoides Chronica . More than 100 cases have been reported so far, however there's still a controversy whether to consider Nekam's disease as a separate entity or as a part of a disease spectrum like Lichen Planus, Lichen Simplex Chronicus or Lupus Erythmatosus[6]. Histologically, there can be hyperkeratosis, follicular parakeratosis, alternating acanthosis and epidermal atrophy. Interface dermatitis along with negative immunofluorescence is present[2,6]. The following three cases describe typical features of the disease in centrofacial distribution and in members of the same family with autosomal recessive pattern of inheritance.

Case 1

A 13-year-old boy, student of 8th grade, resident of Mianwali presented with erythematous papular eruption in seborrheic distribution of face, trunk and limbs since early childhood. It was not associated with itching, photosensitivity or

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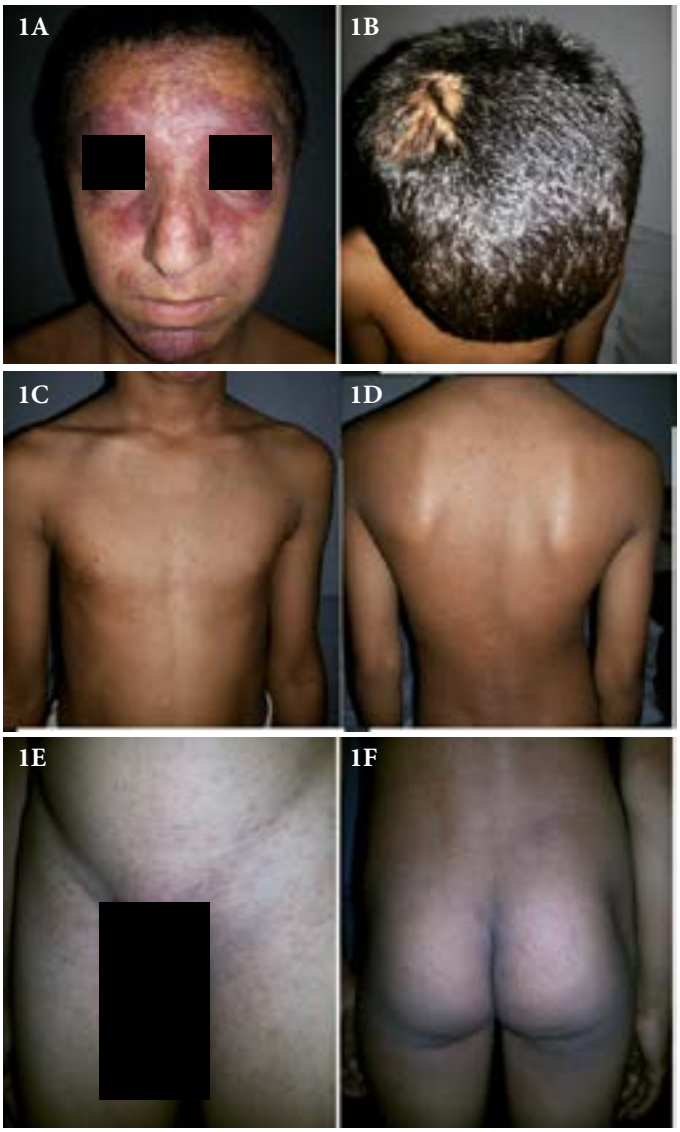

Figure 1: Erythematous papules and plaques confluent in seborrheic region of face. B. Alopecia scalp. C&D. Discrete violaceous papules on Chest and trunk E. Papules with scale in groin F. Papules buttock and back

fever. Lesions worsened during summer and remitted in winter. On cutaneous examination there were multiple violaceous papules with rough glistening surface coalescing over the forehead in a reticulate pattern and formed plaques on nose, periorbital area, cheeks, and chin (Fig 1A). Multiple skin colored to violaceous hyperkeratotic discrete papules, were disseminated over chest, abdomen, upper arms, back, thighs and lower limb. Papules were denser and more grouped in groin, genital area and buttocks (Fig 1C&1F). Hyperpigmented plaques were present in both axillae. There was loss of eye lashes and eye brows. He had three well demarcated round to oval hair loss patches on the scalp along with seborrheic dermatitis like scales all over the scalp (Fig 1B). Nails, palms, soles, post auricular region, bulbar and genital mucosa were normal.

CAPSULE SUMMARY

Three cases of rare Nekam's disease are reported with unusual centro-facial distribution in members of the same family with autosomal recessive pattern of inheritance.

Case 2

A 14-year-old girl, student of 8th grade and resident of Mianwali presented from same family presented with rash on face, scalp, trunk and flexures since childhood. Systemic enquiry was unremarkable. Rash aggravated in summer while spontaneously recovered to near normal in winter. On examination she had multiple violaceous papules on face, in a reticulate manner on forehead and coalescing to form plaques over bridge of nose, periorbital area and chin (Fig 2A). She also had similar papules along hairline (Fig 2C). Similar skin colored to violaceous papules discrete and forming a reticulate network were present on her back and flexures (Fig 2 D&F). Seborrheic dermatitis like scales were present on scalp (Fig 2B).

Case 3

A 3-year-old boy, resident of Mianwali presented with rash on his face, trunk, buttocks and flexures since the age of 2 months. He also had history of exaggeration of rash in summer and resolution in winter. On examination he had multiple violaceous papules on the face predominantly in Centro-

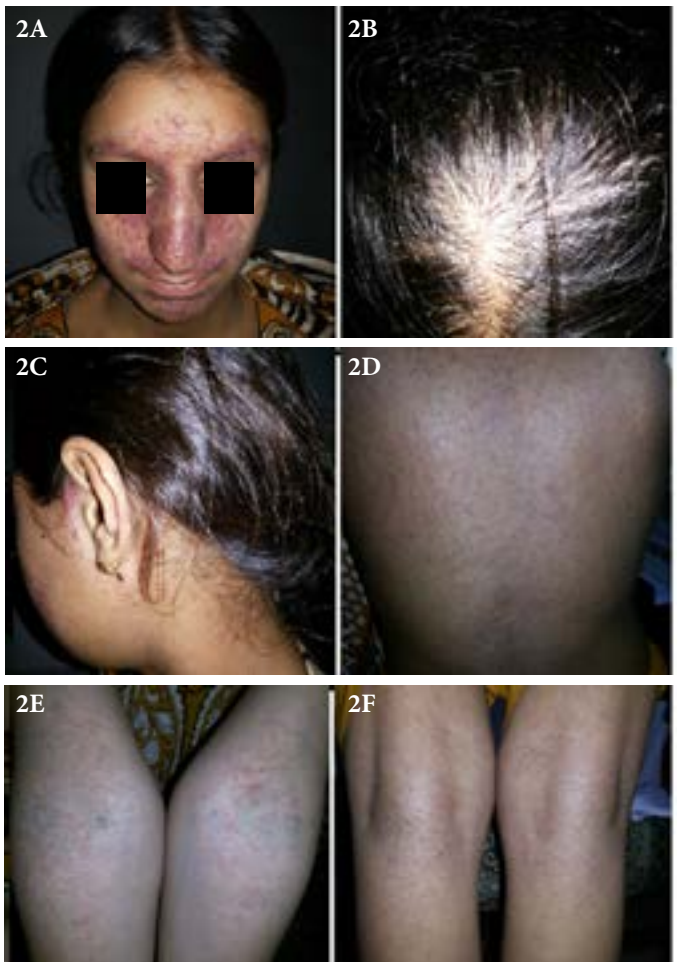

Figure 2: (A) Papules in seborrheic distribution face. (B) Seborrheic dermatitis scalp. (C) Papules along hair line. (D) Papules on back. (E) Papules in elbow flexure. (F) Papules back

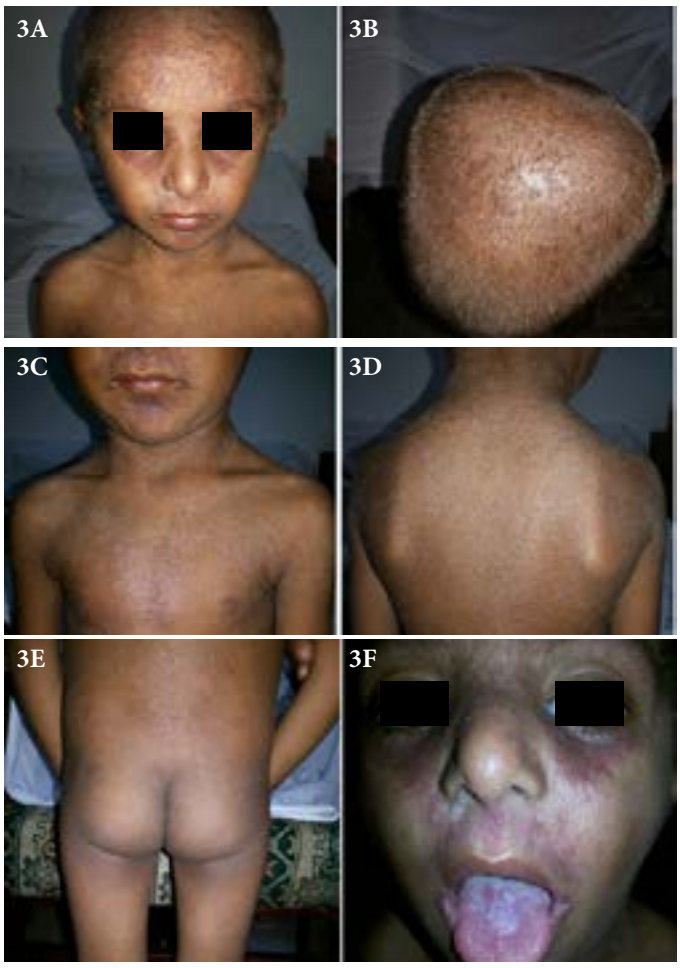

Figure 3: (A) Face. (B) Seborrhoeic dermatitis scalp. (C) Discrete papules on chest. (D) Papules back. (E) Keratotic papules back. (F) Angular chelitus

facial distribution i.e. involving forehead, periorbital area, and bridge of nose, upper lip and chin [Fig 3A). Similar skin colored to violaceous hyperkeratotic papules was present on the chest, back and buttocks with relative sparing of abdomen in a reticulate pattern [Fig 3C&E). He also had involvement of flexures, axillae and groins. There were abundant scales on the scalp (Fig 3B). He also had white plaques on his tongue along with apthous ulcers and angular cheilitis (Fig 3F). There was no involvement of nails, palms and soles.

Differentials of Lichen Planus, Lupus Erythmatosus, Keratosis Lichenoides Chronica (Nekam's disease), and Dariers disease were considered in view and investigated accordingly. Blood complete picture, biochemistries and ANA of all 03 cases were normal. Skin biopsy on histopathology showed hyperkeratosis with parakeratosis, alternate areas of acanthosis and epidermal atrophy. Granular layer was thickened, whereas basal cell vacuolar degeneration was seen at dermoepidermal junction along with mixed cellular infiltrate comprising of lymphocytes, plasma cells and histiocytes. Follicular plugging was also noted along with dilated vessels which confirm the diagnosis of Nekam's disease.

All of above described cases are siblings from a single family. The pedigree is consistent with autosomal recessive inheritance due to presence of affected cases in both genders and skip generations.

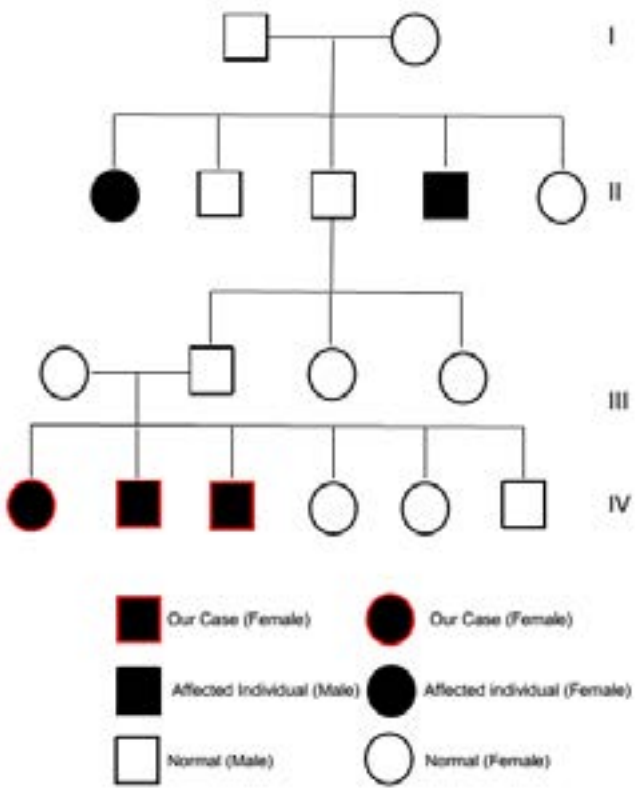

Figure 4: Pedigree showing Autosomal Recessive Pattern of Inheritance

DISCUSSION

Nekam's disease is a reticulate dermatosis that primarily affects adolescents and the young adults, with a slight male predominance (1.35:1)[4]. Rosacea like facial seborrheic dermatitis is present in 75% of cases[3,7]. In 50% of the cases, aphthous ulcers can be found on oral mucosa[8]. Ocular mucosa can also be involved in the form of conjunctivitis, uveitis, iridocyclitis and blepharitis[9]. Forty percent patients can have palmoplantar keratoderma[4,10]. Nail dystrophy is present in 30% of the cases with the commonest nail findings include paronychia and longitudinal ridging[11]. Similarly, hyperkeratotic papules can also be found in males over the scrotum and penis[1]. Multiple aphthous ulcers on the oral mucosa and violaceous edged erosions on the inner side of the prepuce have been reported[12]. In above mentioned cases we had violaceous papules in reticulate pattern on face, trunk and flexures, along with genital involvement in both male patients and seborrhea in all three cases. Oral mucosal was involved in one case. In contrast to two cases described with improvement in summer, these 3 cases reported worsening in summer[13]. It has been observed that Nekam's disease is strongly associated with Glomerulonephritis and Lymphoproliferative disorders8 whereas on histopathological findings of porokeratosis and amyloid deposition can be seen[14]. Nekam's disease has a prolonged, chronic & progressive course of illness and is usually resistant to treatment. Successful treatment has been done using phototherapy without[15,16] or with Acitretin[17]; Etretinate[18] and photodynamic therapy[19].

Our cases responded well to topical steroids and emollients with improvement in erythema and number of papules. They were counselled about hydration and sun protective measures.

CONCLUSION

Nekam's disease is a rare condition. It should be ruled out in patients with apthous ulcers, seborrheic dermatitis or lichen planus. The evaluation of similar cases can lead to diagnosis of more cases of this disorder.

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