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C O N T E N T S

EDITORIAL

Management of autoimmune urticaria

Arun C. Inamadar, Aparna Palit 89

VIEW POINT

Cosmetic dermatology versus cosmetology: A misnomer in need of urgent correction

Shyam B. Verma, Zoe D. Draelos 92

REVIEW ARTICLE

Psoriasiform dermatoses

Virendra N. Sehgal, Sunil Dogra, Govind Srivastava, Ashok K. Aggarwal 94



ORIGINAL ARTICLES

A study of allergen-specific IgE antibodies in Indian patients of atopic dermatitis

V. K. Somani 100

Chronic idiopathic urticaria: Comparison of clinical features with positive autologous serum skin test

George Mamatha, C. Balachandran, Prabhu Smitha 105



Autologous serum therapy in chronic urticaria: Old wine in a new bottle

A. K. Bajaj, Abir Saraswat, Amitabh Upadhyay, Rajetha Damisetty, Sandipan Dhar 109

Use of patch testing for identifying allergen causing chronic urticaria

Ashimav Deb Sharma 114

Vitiligoid lichen sclerosis: A reappraisal

Venkat Ratnam Attali, Sasi Kiran Attali 118



BRIEF REPORTS

Activated charcoal and baking soda to reduce odor associated with extensive blistering disorders

Arun Chakravarthi, C. R. Srinivas, Anil C. Mathew 122



Nevus of Ota: A series of 15 cases

Shanmuga Sekar, Maria Kuruvila, Harsha S. Pai 125



Premature ovarian failure due to cyclophosphamide: A report of four cases in dermatology practice

Vikrant A. Saoji 128

CASE REPORTS

Hand, foot and mouth disease in Nagpur

Vikrant A. Saoji 133



Non-familial multiple keratoacanthomas in a 70 year-old long-term non-progressor HIV-seropositive man

Hemanta Kumar Kar, Sunil T. Sabhnani, R. K. Gautam, P. K. Sharma, Kalpana Solanki, Meenakshi Bhardwaj 136



Late onset isotretinoin resistant acne conglobata in a patient with acromegaly

Kapil Jain, V. K. Jain, Kamal Aggarwal, Anu Bansal 139



Familial dyskeratotic comedones

M. Sendhil Kumaran, Divya Appachu, Elizabeth Jayaseelan 142



Nasal NK/T cell lymphoma presenting as a lethal midline granuloma

Vandana Mehta, C. Balachandran, Sudha Bhat, V. Geetha, Donald Fernandes



145

Childhood sclerodermatomyositis with generalized morphea

Girishkumar R. Ambade, Rachita S. Dhurat, Nitin Lade, Hemangi R. Jerajani.....



148

Subcutaneous panniculitis-like T-cell cutaneous lymphoma

Avninder Singh, Joginder Kumar, Sujala Kapur, V. Ramesh.....



151

LETTERS TO EDITOR

Using a submersible pump to clean large areas of the body with antiseptics

C. R. Srinivas



154

Peutz-Jeghers syndrome with prominent palmoplantar pigmentation

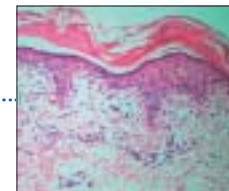
K. N. Shivaswamy, A. L. Shyamprasad, T. K. Sumathi, C. Ranganathan



154

Stratum corneum findings as clues to histological diagnosis of pityriasis lichenoides chronica

Rajiv Joshi



156

Author's reply

S. Pradeep Nair

157

Omalizumab in severe chronic urticaria

K. V. Godse.....

157

Hypothesis: The potential utility of topical eflornithine against cutaneous leishmaniasis

M. R. Namazi

158

Nodular melanoma in a skin graft site scar

A. Gnaneshwar Rao, Kamal K. Jhamnani, Chandana Konda



159

Palatal involvement in lepromatous leprosy

A. Gnaneshwar Rao, Chandana Konda, Kamal Jhamnani..... 161



Unilateral nevoid telangiectasia with no estrogen and progesterone receptors in a pediatric patient

F. Sule Afsar, Ragip Ortac, Gulden Diniz 163



Eruptive lichen planus in a child with celiac disease

Dipankar De, Amrinder J. Kanwar..... 164



Xerosis and pityriasis alba-like changes associated with zonisamide

Feroze Kaliyadan, Jayasree Manoj, S. Venkitakrishnan..... 165

Treatment of actinomycetoma with combination of rifampicin and co-trimoxazole

Rajiv Joshi 166



Author's reply

M. Ramam, Radhakrishna Bhat, Taru Garg, Vinod K. Sharma, R. Ray, M. K. Singh, U. Banerjee, C. Rajendran 168

Vitiligo, psoriasis and imiquimod: Fitting all into the same pathway

Bell Raj Eapen 169

Author's reply

Engin Şenel, Deniz Seçkin 169

Multiple dermatofibromas on face treated with carbon dioxide laser: The importance of laser parameters

Kabir Sardana, Vijay K. Garg 170

Author's reply

D. S. Krupa Shankar, A. Kushalappa, K. S. Uma, Anjay A. Pai..... 170

Alopecia areata progressing to totalis/universalis in non-insulin dependent diabetes mellitus (type II): Failure of dexamethasone-cyclophosphamide pulse therapy

Virendra N. Sehgal, Sambit N. Bhattacharya, Sonal Sharma, Govind Srivastava, Ashok K. Aggarwal 171



Subungual exostosis

Kamal Aggarwal, Sanjeev Gupta, Vijay Kumar Jain, Amit Mital, Sunita Gupta..... 173

Clinicohistopathological correlation of leprosy

Amrish N. Pandya, Hemali J. Tailor 174

RESIDENT'S PAGE

Dermatographism

Dipti Bhute, Bhavana Doshi, Sushil Pande, Sunanda Mahajan, Vidya Kharkar 177

FOCUS

Mycophenolate mofetil

Amar Surjushe, D. G. Saple 180

QUIZ

Multiple papules on the vulva

G. Raghu Rama Rao, R. Radha Rani, A. Amareswar, P. V. Krishnam
Raju, P. Raja Kumari, Y. Hari Kishan Kumar 185



E-IDVL

Net Study

Oral isotretinoin is as effective as a combination of oral isotretinoin and topical anti-acne agents in nodulocystic acne

Rajeev Dhir, Neetu P. Gehi, Reetu Agarwal, Yuvraj E. More 187

Net Case

Cutaneous diphtheria masquerading as a sexually transmitted disease

T. P. Vetrichevvel, Gajanan A. Pise, Kishan Kumar Agrawal,
Devinder Mohan Thappa 187



Net Letters

Patch test in Behcet's disease

Ülker Gül, Müzeyyen Gönül, Seray Külcü Çakmak, Arzu Kılıç 187

Cerebriform elephantiasis of the vulva following tuberculous lymphadenitis

Surajit Nayak, Basanti Acharjya, Basanti Devi, Satyadarshi Pattnaik,
Manoj Kumar Patra 188



Net Quiz

Vesicles on the tongue

Saurabh Agarwal, Krishna Gopal, Binay Kumar 188



Familial dyskeratotic comedones

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ABSTRACT

Familial dyskeratotic comedones (FDC) is a rare autosomal dominant inherited condition, characterized by widespread, symmetrically scattered, comedone-like, hyperkeratotic papules, which are cosmetically unappealing. These lesions appear around puberty and show a predilection to involve the trunk, arms and face. The lesions are asymptomatic and gradually worsen with time. Histology shows invagination of the epidermis with a lamellar keratinous plug and focal evidence of dyskeratosis. This condition is generally refractory to therapy. We report here two cases with this rare disorder who had a strong familial history of the same disorder.

Key Words: Autosomal dominant, Comedone, Dyskeratosis, Familial dyskeratotic comedone

INTRODUCTION

Familial dyskeratotic comedones (FDC) is a rare asymptomatic autosomal dominant condition with distinctive clinical and histopathological features. Ever since its preliminary report by Rodin *et al.*^[1] in 1967, only a few cases have been reported in literature around the world. It is clinically characterized by scattered, hyperkeratotic comedone-like papules.^[2] Histopathology shows crater-like invaginations filled with keratinous material and evidence of dyskeratosis.^[3] To the best of our knowledge, only 15 patients from seven families have been reported in literature^[1-8] and there is no case reported from India. We report here two families from India with this rare disorder.

CASE REPORTS

Case 1

A 21 year-old female presented with an eight year-old history of multiple, asymptomatic widespread, hyperkeratotic papules. Her lesions initially appeared over the chest and subsequently spread to involve all areas of the body except the scalp, legs, mucosa, palms and soles. Apart from these skin lesions, multiple pock-like scars were

seen over the face and the back. Removal of a keratinous plug from one of the papules revealed a minimal bleeding crater. There was no evidence of acne. The lesions started as pinpoint dark papules, which increased in number and extent of involvement as the patient grew older. When fully formed, the lesions measured around 0.5 to 1 cm in size [Figure 1].

A few of the keratotic papules formed painful, inflamed swellings over the back. The patient had not received any treatment and was in good health and otherwise normal. A family history of similar lesions, that had started around the age of 12 years, was noted in her mother, two younger sisters and a maternal uncle [Figure 2].

Skin biopsy from the hyperkeratotic lesions revealed a crater-like invagination filled with lamellar keratinous material [Figure 3]. Foci of dyskeratosis were seen and a few of the dilated follicles showed colonization with budding yeast cells. Skin biopsies from the patient's mother and sister revealed similar histopathological findings. The patient was treated with oral itraconazole 200 mg/day for one week resulting in a decrease in the number of keratotic papules and inflamed swellings.

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Figure 1: Pock-like scars and comedones - case 1

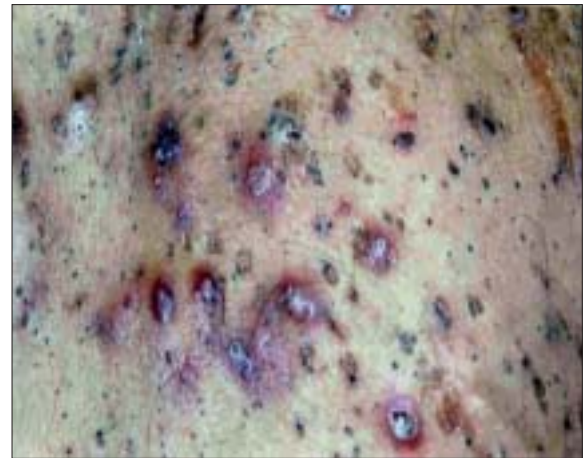


Figure 4: Multiple inflamed nodules and comedones over back

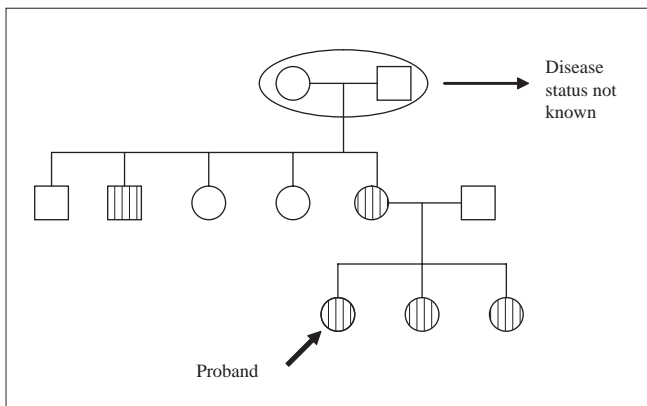


Figure 2: Pedigree chart-Case 1

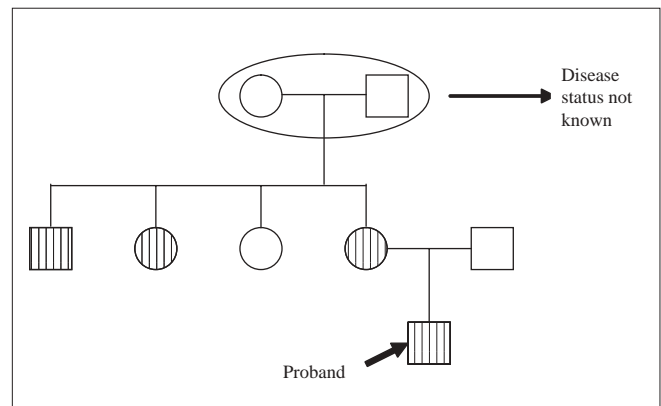


Figure 5: Pedigree chart-Case 2

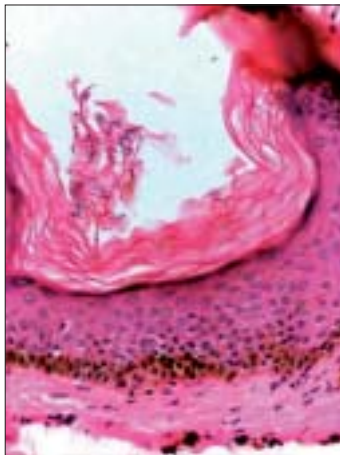


Figure 3: Dilated follicular ostia filled with lamellar keratin with yeasts (H and E stain, 20x)

Case 2

A 30 year-old male presented with hyperkeratotic papules (similar to case 1) over the trunk, upper limbs, face and thighs which initially appeared around 15 years of age over upper limbs and increased progressively since then. He also gave a history of tender swelling over the back [Figure 4],

which appeared when the keratotic papules increased in size. Case 2 had more lesions than did case 1. He also had large pock-like scars over his face. Physical examination aside from the skin lesions was normal. He had been treated with isotretinoin 20 mg daily for five months before he came to us with no improvement. Family history of similar lesions was seen in the mother, maternal uncle and aunt [Figure 5]. Histopathology showed similar changes as those in case 1.

DISCUSSION

Carneiro *et al.*^[3] proposed the term FDC when he first described a family of four members affected with this rare entity, which was based on the following distinctive features:

1. Lesions clinically resembling comedones
2. Occurrence in some family members
3. Presence of dyskeratotic changes on histological examination

McKusick^[2] recently classified the disease as autosomal dominant (McK 120450). As the condition is asymptomatic,

it may only be seen as an incidental finding. Complete family history (all family members) should be taken whenever possible.

The lesions usually appear around puberty and show a predilection for the trunk, arms, legs, face and shaft of the penis, sparing the glans, palms and soles.^[2] Histologically, it is characterized by dyskeratosis and invaginations into the dermis, occasionally acantholysis may be seen. Dyskeratosis may not be seen in all patients.^[9] Electron microscopy in FDC shows a decreased number of desmosomal attachments within the stratum malpighii.^[5]

Both our index cases had clinical and histopathological features which correlated well with the diagnosis of FDC. However, there was one peculiar finding in both our patients; a few of the follicular ostia were filled with budding yeasts cells. It is not known how these yeasts were contributing to the pathogenesis of the disease. Culture or scrapping for the fungus was not attempted as it is well known that *P. ovale* colonizes in the pilosebaceous ducts.^[10] Nearly half of the cases reported in literature had concomitant acne and only one was reported to have severe acne cysts. Both our patients gave history of multiple inflamed acne cysts over the back, most of which led to secondary scarring.

Kyrle's disease, reactive perforating collagenosis, keratosis pilaris, perforating folliculitis, nevus comedonicus, acne vulgaris and Darier disease must be considered in the differential diagnosis.^[1,3,6] Treatment has always been unrewarding. Various treatment modalities including topical retinoids and oral isotretinoin have proved to be ineffective.^[7] The pathophysiological process in FDC could be different from that of normal comedones in acne, thus explaining its lack of response to retinoid treatment. However, frequent sun exposure^[5] and carbon dioxide laser^[5] have shown promising results. Our first patient has shown good results with itraconazole and has been on regular follow-up with us. We found that when the patient was put on itraconazole pulse therapy, a dramatic response

in the form of healing of the inflamed lesions along with the absence of new lesions was noted. The exact mode of action of itraconazole remains unknown; it probably decreased the local yeast colonization.

FDC is usually asymptomatic, hence, it may go unseen. However, in patients where the face is also involved, it could lead to many social problems especially in the female population. Data regarding the long-term follow-up of these patients are lacking. The prognosis is generally better as the patients are otherwise healthy.

This report highlights both the efficacy of itraconazole and reports two cases of this rare genodermatosis, which to the best of our knowledge, is the first report from India.

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