



**ORIGINAL RESEARCH PAPER**

**Dermatology**

**FAMILIAL DYSKERATOTIC COMEDONES – A RARE CASE REPORT**

**KEY WORDS:**  
Dyskeratosis, grouped comedones, desmosomes.

<b>Sudheer Kumar S*</b>	Junior Resident, Department of DVL, Sri Venkateswara Medical College, Tirupati. *Corresponding Author
<b>Ram Kumar KR</b>	Junior Resident, Department of DVL, Sri Venkateswara Medical College, Tirupati.
<b>Chenchaiah V</b>	Junior Resident, Department of DVL, Sri Venkateswara Medical College, Tirupati.
<b>Venkata Ramana P</b>	Professor, Department of DVL, Sri Venkateswara Medical College, Tirupati.

**ABSTRACT**

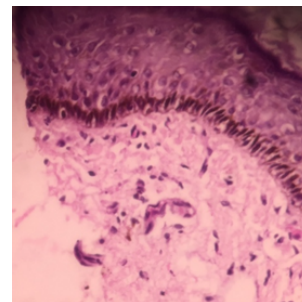
Familial dyskeratotic comedones (FDC) is a rare autosomal dominant inherited condition, characterized by widespread, symmetrically scattered, comedone-like, hyperkeratotic papules, which are cosmetically unappealing. Lesions show a predilection to involve the trunk, arms and face and appear around puberty. The lesions are usually asymptomatic and worsen gradually 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 a case with this rare disorder.

**INTRODUCTION**

Familial dyskeratotic comedones (FDC) is an uncommon autosomal dominant disorder with distinctive clinical features characterized by discrete and disseminated numerous comedones and hyperkeratotic papules which are refractory to treatment. On histopathology, it shows crater-like invaginations filled with keratinous material and evidence of dyskeratosis (1).

**CASE REPORT**

A 40 year old male presented with multiple asymptomatic and few tender skin color nodules and dark patches of varying sizes ( 1x2 to 2x3 cm) predominantly over trunk, upper arms and thighs. Initially started over both legs at the age of 30 years which gradually increased in size and number and subsequently spread to involve trunk and upper arms. History of similar lesions present in his father. Numerous wide spread comedones varying in size from 0.3 to 0.5 cm over the trunk and thighs along with multiple pock like scars on thighs. Face, mucosa, palms & soles are spared. Systemic examination and routine hematological investigations were normal. Histopathology revealed plugs of keratin



**Fig-4**

Plugs of keratin (Fig-3) in follicles and (Fig-4) dyskeratotic cells

**DISCUSSION**

FDC has characteristic features of clinical lesions resembling comedones, positive family history, dyskeratotic changes on HPE. Acantholysis may also be seen sometimes. Usually presents in adolescence. The lesions start as pinpoint dark papules favouring trunk, arms, legs, face and shaft of the penis (sparing the glans), oral mucosa, palms, and soles. EM reveals decreased number of desmosomal attachments within the stratum malpighii (2),(3). Differential diagnosis include Comedonal Darriers, Kyrles disease, keratosis pilaris, Nevus comedonicus and Acne vulgaris(2),(3). The classical clinical features, positive family history, and histopathology confirmed diagnosis.

**CONCLUSION**

We report this case because of its rarity, sparing of face and with large scars and nodular lesions which was not reported in previous case reports.

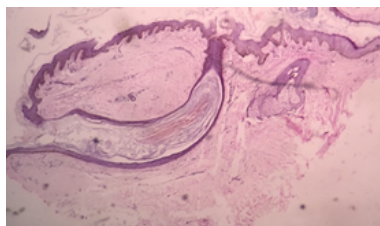
**REFERENCES:**

1. Familial dyskeratotic comedones Kumaran M S, Appachu D, Jayaseelan E - Indian J Dermatol Venereol Leprol [Internet]. [cited 2019 Jun 9]. Available from: <http://www.ijdv.com/article.asp?issn=0378-6323;ye ar=2008;v olume =74;issue=2;spage=142;epage=144;a ulast=Kumaran>
2. Familial dyskeratotic comedones: A rare entity Maddala RR, Ghorpade A, Polavarpu M, Adulkar SA, Das M - Indian Dermatol Online J [Internet]. [cited 2019 Jun 9]. Available from: <http://www.idoj.in/article.asp?issn=2229-5178;year=2016;volume=7;issue=1;spage=46;epage=48;a ulast=Maddala>
3. Familial Disseminated Comedones without Dyskeratosis: Report of an Affected Family and Review of the Literature, Mao-Jie Cheng a Wen-Chieh Chen b Rudolf Happle c Zhi-Qiang Song a, Dermatology 2014;228:303-306



(a) in follicles and few (b) dyskeratotic cells in malpighi layer.

**Multiple Pock Like Scars With Multiple Comedones And Nodules**



**Fig-3**