DOWLING-DEGOS DISEASE, HIDRADENITIS SUPPURATIVA AND EPIDERMOID CYST - A RARE CASE REPORT

Rakesh1, Savita Agrawal1, Manisha Nijhawan3

12nd Year Postgraduate Resident, Department of Skin & V.D., Mahatma Gandhi Medical College & Hospital, Jaipur.
2Assistant Professor, Department of Skin & V.D., Mahatma Gandhi Medical College & Hospital, Jaipur.
3Professor & HOD, Department of Skin & V.D., Mahatma Gandhi Medical College & Hospital, Jaipur.

ABSTRACT

BACKGROUND
A 23-year-old female patient presented with flexural pigmentary lesions, comedones and pitted scars over face suggestive of Dowling-Degos disease of 10 years duration. Patient also had multiple, painful, relapsing, nodules with sinuses discharging pus and scars in apocrine gland bearing region suggestive of hidradenitis suppurativa since past 2 years and single painless cystic lesion over labia majora suggestive of epidermoid cyst for past 1 year 2 months. We are reporting a rare clinical case with all the three diseases suggestive of common defect in follicular epithelial proliferation, which is not reported in Indian literature till date.

KEYWORDS
Dowling-Degos Disease, Hidradenitis Suppurativa, Epidermoid Cyst.


BACKGROUND
Dowling-Degos disease is a rare genetic disease of the skin that presents with asymptomatic hyperpigmented macules in the skin folds with prominent comedone like lesions and pitted scars.1,2,3,4 It can either be inherited from one parent (Autosomal dominant) or appear without a family history (Sporadic) with female predominance.5,6 Other associated findings include hidradenitis suppurativa,7,8 keratocanthoma,9 epidermoid cyst10 and squamous cell carcinoma.10

Case Report
A 23-year-old female patient presented with multiple, small hyperpigmented macules over face and flexures for past 10 years. Initially, lesions appeared over the face, gradually progressing to involve neck [Figure 1], axilla, antecubital fossa and groin. Scattered comedo-like lesions (Dark dot follicles) were seen over the axilla [Figure 2], abdomen, buttocks, thighs and anogenital area for past 7 years. The patient had multiple discrete pitted scars over the face mainly over the perioral region [Figure 3]. The lesions were asymptomatic, but progressive in nature. She developed multiple, painful, tender nodular lesions with multiple sinuses discharging pus over the buttocks [Figure 4], inner aspect of upper thigh and anogenital region on and off since past 2 years. She noticed a painless, cystic swelling over right portion of labia majora [Figure 5] for past 1 year 2 months. There was a history of similar pigmented lesion in her mother. The patient was born of a non-consanguineous marriage. Her general physical examination and systemic examinations were normal.

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Corresponding Author:
Dr. Rakesh,
Room No. 312, Type 4B P.G. Hostel,
Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.
E-mail: iangradrrakesh@yahoo.in, m.nijhawan12@gmail.com

Figure 1. Hyperpigmented Macule over Neck

Figure 2. Comedones and Hyperpigmented Macules over Axilla

Figure 3. Scar and Hyperpigmented Macules over Chin


