SHORT COMMUNICATIONS

DOWLING-DEGOS DISEASE

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Four cases of Dowling-Degos disease with classical reticulate, flexural pigmentation are described. One of the four patients had milia, while all of them had perioral pitted scars.

Key Words : Reticulate pigmentation, Dowling-Degos disease, Flexures

Introduction

Reticulate pigmentary disorders as a group are characterised by hyperpigmented macules of various intensities of brown or black.[1] In many disorders they are freckle like and angulated and tend to join at their margins to form a reticulate pattern. A second distinct element in some conditions, but not in others, is the presence of hypopigmented macules scattered between the hyperpigmented lesions. Dowling-Degos disease (DDD) is a type of reticulate pigmentary disorder in which hyperpigmented macules are found in the flexures. It was first established by Wilson-Jones and Grice,[2] though it was described as early as 1938. Few reports have appeared in the Indian literature.[3-5] We describe four cases from four different families with other affected members who were also examined.

Case Reports

Case 1: A 25-year-old woman noticed progressive pigmentation of the neck, upper chest, back, cubital fossae and axillae since the age of 19 years. She also noticed appearance of multiple, pin-head sized white papules on her right eyelids, sides of the nose and cheeks for the last 4 years. She had no similarily affected family member. On examination reticulate pigmentation was present in the axillae, cubital fossae, on the sides of the neck, upper chest and back. There were multiple milia on the eyelids, nose and cheeks and pitted scars in perioral area (Fig.1).

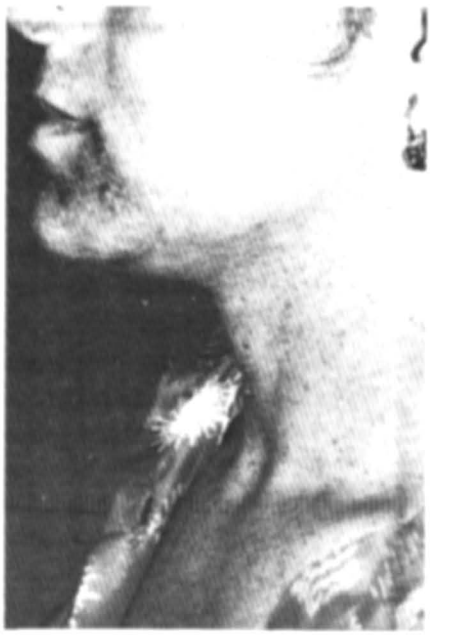

Fig. 1. Pigmentation of the neck and pits in perioral area (case 1).

Histological examination of the pigmented area showed, a normal epidermis

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with delicate, melanized, epidermal downgrowths characteristic of DDD (Fig.2),

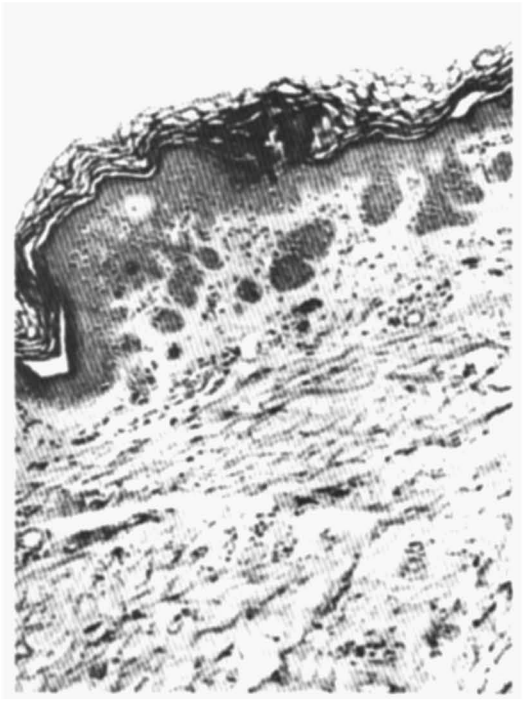

Fig. 2. Delicate, antler-like downgrowths with hypermelanization at tips (H&E x 200).

that from the milia showed normal epidermis with cystic structures lined by squamous epithelium, connected to hair follicles in the dermis.

Case 2: A 16-year-old girl noticed multiple pigmented spots on her wrist, cubital fossae, popliteal fossae and axillae at the age of 12 years. They increased in number and spread to adjacent areas. Examination revealed multiple, angulated, hyperpigmented macules in all flexures, on the sides of neck, face and temples. There were multiple pitted scars in the perioral area. Her mother, maternal aunt and grandfather were also similarily affected. Histological findings were similar to case 1.

Case 3: A 22-year-old man had multiple, small, hyperpigmented macules in the flexures, on sides of the neck, face and forehead since the age of 10 years. There were multiple perioral pits. Histological findings were similar to case 1. His mother and maternal aunt were also similraily affected.

Case 4: A 14-year-old-girl developed reticulate pigmentation at the age of 11 years at similar sites as case 1. Histological findings were similar to case 1. Her mother and elder sister were suffering from similar problem.

Discussion

Dowling-Degos disease, an autosomal dominant genodermatosis, has been described rarely in Indian literature.[3-5] It is characterised by reticulate pigmentation of the flexures, comedo-like papules and perioral pitted scars.[2]

Our patients had features similar to those described in the literature.[2] All except case I had family members affected demonstrating an autosomal dominant pattern of inheritance. One male was found to be equally affected though the disease has been reported to be milder in nature in men.[6] The distribution of pigmentation, presence of perioral pitted scars in all patients was similar to earlier reports. Only 1 patient had epidermoid cysts which have been reported by others.[6] None of our patients had seborrhoeic keratosis, follicular hyperkeratoses, dry skin, comedo-like-lesions as have been described earlier.[6]

References

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MR BOIL'S FAMILY TREE

Mr Boil has a rather unique family tree
To describe it in detail I'm afraid would be quite beyond me.
I make therefore, a clinical reference to some of his clan
To offend the rest I assure you is not part of my plan!

His children are curiously named EB & Impetigo Bullous
The birth of the first was traumatic; the other is by nature infectious.
Then there is the toddler herpes who had the pox the other day,
I'm told he broke into bubbles while eating his curds & whey!!

Uncle pemphigus one may say is a touch 'FLACCID'
Being on steroids, he has become quite PLACCID,
Aunt pompholyx- she's the tyrannic one in the PHYLUM,
Rumour has it Saddam has just granted her 'ASYLUM'!

Grandpa pemphigoid tends to get rather 'TENSE'now a days
& is constantly seen trying to get the children to mend their ways;
Grandma burns spilt hot water on to her 'Eczema' yesterday
& is seen spending her Sunday 'Boiling-over' in sick bay!

Father EMF developed a drug reaction while praying last year
& ever since, has been drowning his sorrows in barrels of beer.
Cousin DH-he's the one in the family who loves to crash parties
& develops this 'ITCH' every time he downs his 'Chappaties'!!
Mr Boil it must suffice to say has an effervescent family tree
Dapsone, retinoids & steroids do visit him rather frequently
His appearances are usually dramatic & cause great disdain
Bulla! A good concept of this tree my friends, will not be in vain!

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