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Case Report

Cutis verticis gyrata and leonine face in a patient with Darier disease: A case report and review of the literature

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Abstract

Cutis verticis gyrata is classified into primary and secondary types. It can be seen in association with chronic inflammatory dermatologic diseases, tumors, and chromosomal and inherited disorders. Here, we describe a patient of Darier's disease presenting with cutis verticis gyrata involving scalp expanding to his forehead leading to leonine face.

Key words
Cutis verticis gyrata, Darier's disease, leonine face.

Introduction

Cutis verticis gyrata (CVG) is characterized by development of gyriform furrows and ridges on scalp mostly in vertical direction from vertex to occiput area. Furrows occasionally involve occipital area in a transversal direction. CVG may occur as both congenital and acquired disorder. Number of the folds varies between 2 to 20 with mean number of 14. Width of each fold ranges from 0.75 cm to 4 cm and depth of each group ranges from 3 to 10 cm with average being 1 cm^2.[2, 1]

Darier disease is an autosomal dominant disease due to mutations in ATP2-A2-gene that leads to abnormal keratinization. The disease is characterized by involvement of skin, mucous membranes and nail. Skin lesions manifest as brownish keratotic papules and plaques that predominantly involve seborrheic areas[2].

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Here, we describe a patient with Darier disease with Cutis verticis gyrata in scalp and frontal areas of his face since adolescence.

Case Report

A 67-year-old man who was a known case of Darier disease presented to dermatology clinic because of exacerbation of skin lesions. He also complained of moderate pruritus and malodor of the lesions. He had numerous papules on scalp, face and upper trunk that some of them coalesced and developed widespread plaques and warty mass (**Figure 1**). He also had macerated verrucous plaques in flexural area i.e. axilla and groin. On palmoplantar areas we found verrucous plaques and keratin filled pits. Nail examination showed longitudinal ridging and fissuring with wedge-shaped notch on free edge of some of his fingers and subungual hyperkeratosis.

Scalp examination demonstrated vertical grooves and furrows with soft consistency distributed from vertex toward occipital area (**Figure 2**). Also there were few furrows in frontal region expanding to mid-face that did not

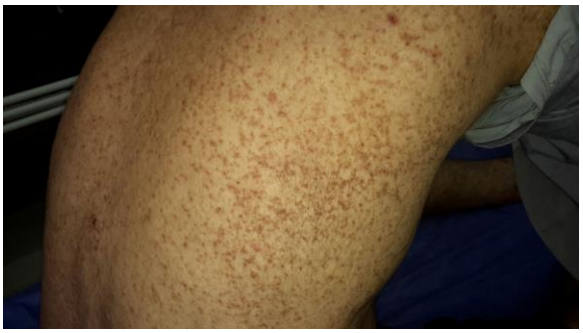

Figure 1 Numerous papules on back that some of them confluent to each other and developed widespread plaques.

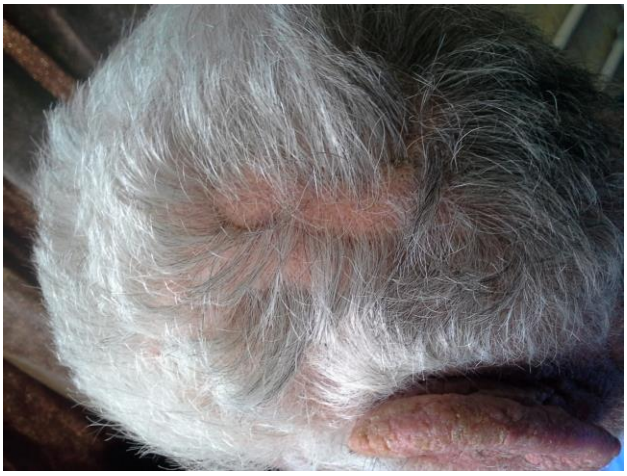

Figure 2 Vertically grooves and furrows with soft consistency distributed from vertex toward occipital area

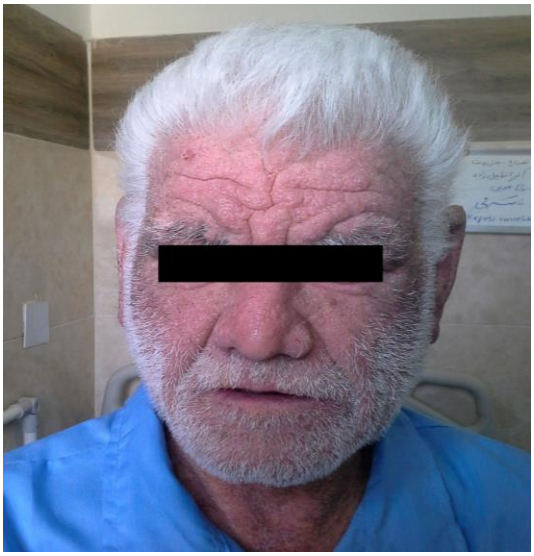

Figure 3 Furrows in frontal region expanding to mid-face.

flat following traction (**Figure 3**). Oral and ophthalmic examination was normal.

Discussion

CVG is classified to primary and secondary types. Primary form is divided to essential and nonessential types.[1] Primary form presents symmetrically in anterior-posterior direction that starts in 90% of the cases before 30 years of age, commonly in puberty. Neurologic abnormalities (mental retardation, seizure, microcephaly, cerebral palsy) and ophthalmic manifestations (cataract, strabismus, blindness, and pigmentosa retinitis) have been recognized in nonessential type of CVG. Conversely in essential type there is no associated disease and it exclusively involves men at puberty.[3]

In secondary form of CVG, the underlying lesion defines the location and distribution of the grooves. Review of the literature reveals that this form may develop after inflammatory dermatosis and internal malignancies as paraneoplastic syndrome (**Table 1**). The most common cause of secondary type is congenital nevus.[4]

CVG usually is asymptomatic, but because of aggregation of skin secretions, occasionally patients complain of pruritus, tenderness, burning sensation, malodor and reduced hair density in folded areas. Skin biopsy of essential primary form reveals connective tissue hypertrophy with adnexal structure hyperplasia. Since CVG is asymptomatic, no treatment is required. In order to decrease foul smell and pruritus, special care by cleaning of the grooves leading to removal of the skin secretion is recommended.[5,6]

Surgical procedures i.e. scalp reduction with or without skin expanders may be considered for cosmetic issues, but mostly have led to unsatisfactory results.[7]

Table 1 Diseases associated with cutis verticis gyrate.

Disease group	*Examples*
Inflammatory dermatosis	Psoriasis [4]
	Eczema
	Keloidal acne
Infectious disease	Impetigo [1]
	Folliculitis
	Erysipelas
	Syphilis
	Tuberculosis
Inherited disease	Turner syndrome [12]
	Noonan syndrome
	Fragile X syndrome [1]
	Klinefelter syndrome
	Hereditary neurologic amyotrophy
	Tuberous sclerosis
	Ehlers-Danlos syndrome
Tumors	Dermatofibroma, Neurofibromatosis
	Lipomatous nevus, Sebaceous nevus, Intradermal nevus
	Lymphangioma
	Histiocytofibroma
	Leukemia
	Cylindroma [4]
Paraneoplastic syndrome	Metastatic carcinoma
	Fallopian tube cancer
	Pituitary tumor
Miscellaneous	Acanthosis nigricans, Acromegaly
	Amyloidosis
	Chronic traction
	Trauma
	Mucinosis
	Myxedema, Pachydermoperiostosis, Hyper-IgE syndrome [12]

Our patient also had skin thickening, furrows and ridges in forehead of his face similar to leonine appearance. To our information, this is the first report of leonine face in Darier disease. Leonine facies classically, has been observed in patients with lepromatous leprosy and with lower frequency in amyloidosis and scleromyxedema. There are reports of developing leonine facies in sarcoidosis and acute myeloid leukemia.[8-11]

Development of leonine face in our patient can be explained by chronicity of the disease leading to numerous infiltrated papules and plaques.

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