Volume 22 Number 12 | December 2016
DOJ 22 (12): 21.

Cutis verticis gyrata

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Abstract

Cutis verticis gyrata that involves only the face is a rare presentation of this even rarer cutaneous anomaly. We present a 61-year-old man, who developed primary essential progressive cutis verticis gyrata of the face.

Case Presentation

PATIENT: 61-year-old man
DURATION: Eight years
DISTRIBUTION: Forehead

HISTORY: A 61-year-old man was seen in the Ulcer Clinic at Bellevue Hospital Center and was referred to Dermatology Clinic for evaluation of a leonine facies. The patient noted progressive wrinkling and thickening of the skin of his forehead over the last thirty-eight years. The change was gradual, and he brought photographs that showed the deepening of the furrows over time. The patient denied any other skin lesions, dysasthesias, or active infections. Past medical history included pre-diabetes, hyperlipidemia, and chronic venous insufficiency.

PHYSICAL EXAMINATION: On the central forehead and glabella were exaggerated skin folds that were interrupted by deep-undulating furrows (**Figure 1**). The scalp was normal. The facial folds gave the appearance of a leonine facies.

LABORATORY DATA: None

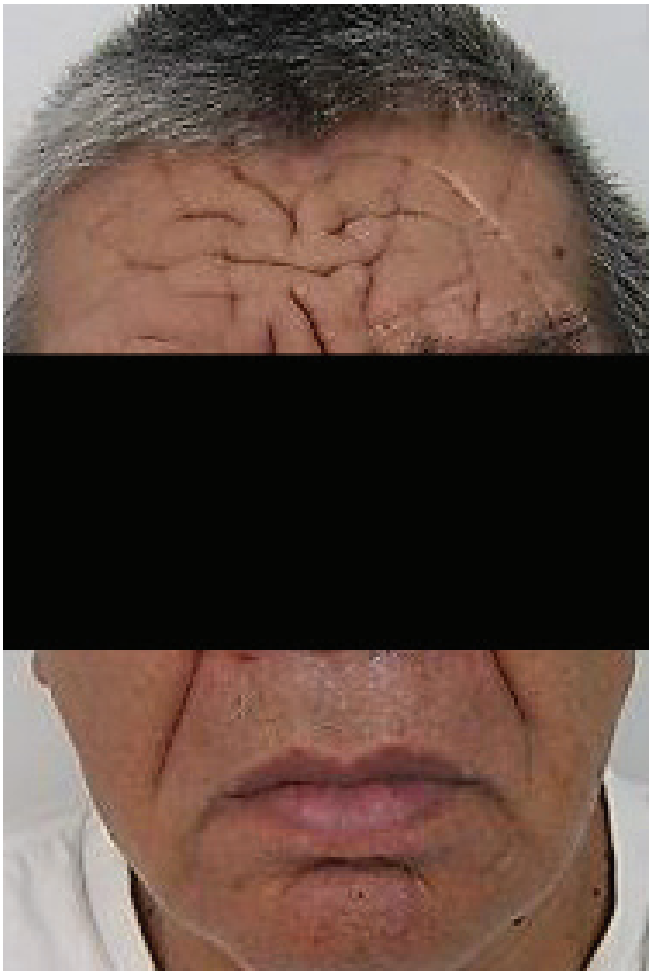

Figure 1. Exagerrated skin folds interrupted by deep-undulating furrows on the central forehead and glabella

HISTOPATHOLOGY: There is a slightly undulating epidermis from which emanates some enlarged sebaceous gland lobules. Occasional thickened collagen bundles are noted (**Figure 2**).

DIAGNOSIS: Cutis verticis gyrata

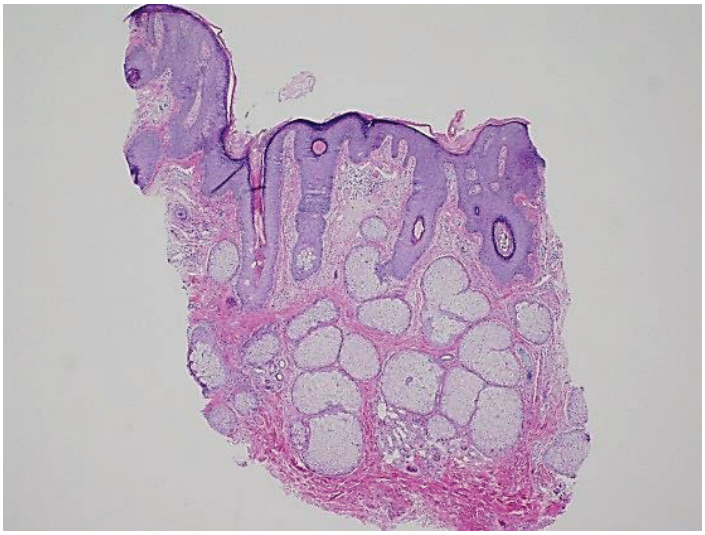

Figure 2. Slightly undulating epidermis from which emanates enlarged sebaceous gland lobules. Occasional thickened collagen bundles are noted

Discussion

Cutis verticis gyrata is a rare, cutaneous condition, which usually affects the scalp with undulating, thick skin folds that are separated by deep sulci, which gives the clinical appearance of brain tissue. Various amounts of cutaneous involvement have been described, with the reported measurements from 2 to 30 folds, each with a width of 0.5 to 2 cm, and furrows that are generally 1 cm in depth [1, 2]. The incidence is higher in men than it is in women, with 1 in 100000 men compared to 0.026 in 100000 women [3].

Cutis verticis gyrata can be a rare primary variant, with no underlying disease, or secondary in the setting of systemic disease [4]. The primary type may be further divided into primary essential and primary non-essential. Primary non-essential is cutis verticis gyrata that is found in association with mental deficiency, cerebral palsy, epilepsy, seizures or ophthalmologic abnormalities [5]. The associated systemic diseases in secondary cutis verticis gyrata include cerebriform intradermal nevi, neurofibromas, acromegaly, myxoedema, Touraine Solente Gole syndrome, leukemia, acanthosis nigricans, and paraneoplastic syndromes [2, 6]. Therefore it is important to take a thorough medical history as cutis verticis gyrata may be the initial sign of one of these underlying diseases [7, 8].

Cutis verticis gyrata is so named since the condition most frequently affects the scalp and specifically the vertex. Few reports exist of this condition affecting only the forehead and glabella [4, 9]. Two case reports of cutis verticis gyrata were associated with hyper-IgE syndrome, and one of these cases affected only the face [4, 10]. The IgE level should be evaluated in our patient. However, associated stigmata of hyper-IgE syndrome, which include atopic dermatitis and recurrent skin and lung infections, were not present in our patient. Owing to this rare presentation of facial involvement only in our patient, it was important to rule out other causes of leonine facies, such as lepromatous leprosy, mycosis fungoides, leishmaniasis, and amyloidosis. For this reason a biopsy specimen that was consistent with cutis verticis gyrata was essential for establishing the diagnosis.

Treatment for cutis verticis gyrata is predominately surgical and is aimed at correction of the cosmetic deficit. Smaller furrows may lend themselves to direct excision and primary closure while larger furrows over more extensive areas have been most amenable to staged procedures with serial tissue expanders and flap advancements [2, 3, 4, 11].

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