Cutis verticis gyrata: three cases illustrating three different etiologies

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Abstract

Cutis Vertcis gyrata is an uncommon neurocutaneous syndrome characterized by excessive growth of the skin of the scalp or the face, forming folds of similar aspect to cerebral cortex gyri. Three categories have been individualized: the primary form, essential or non-essential, and the secondary form.

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Abstract:

Cutis Vertcis gyrata is an uncommon neurocutaneous syndrome characterized by excessive growth of the skin of the scalp or the face, forming folds of similar aspect to cerebral cortex gyri. Three categories have been individualized: the primary form, essential or non-essential, and the secondary form.

Key words: cutis verticis gyrata, neurocutaneous syndrome, folds, hypertrophy

Key Clinical Messages:

- Cutis verticis gyrata is a rare neurocutaneous syndrome with mainly anesthetic impact.

- A rigorous clinical examination with a minimal workup is required to eliminate a secondary or a primary non-essential form.

Introduction:

Cutis verticis gyrata (CVG) is a rare skin disease. affecting 1 of 100,000 males and 0.026 of 100,000 females in the general population [1] CVG is characterized by excessive growth of the skin of the scalp or the face, forming folds of similar aspect to cerebral cortex gyri. Three forms have been individualized: the primary form, essential or non-essential, and the secondary form. The main complaint of the patients is usually cosmetic. The treatment, if requested, is surgical. We report three cases that illustrate one of each of these three categories.

Observations:

Case 1: A 28-year-old male presented with multiple trichilemmal cysts of the scalp. Physical examination revealed, in addition, a thickened scalp with multiple symmetric skin folds from vertex to occipital area (figure 1). The systemic examination did not reveal any neurologic, ophthalmologic, or metabolic anomalies. Laboratory investigations were within the normal range limit. The diagnosis of primary essential CVG was retained and the patient required only excision of the cysts.

Case 2: A 32-year-old male with a history of epilepsy was referred to our consultation for a drug reaction secondary to phenobarbital. On examination, apart from the skin lesions consistent with the drug reaction, the patient had cerebriform folds on his forehead. The furrows were running horizontally and had a width of 1 to 2 cm (figure 2). He didn't have any lesions on the scalp. No other anomalies were found. The diagnosis of primary non-essential CVG associated with epilepsy and involving only the forehead was retained.

Case 3: A 54-year-old female treated for hypertension, dyslipidemia, and a heart rhythm disorder, presented to our department with an occipital tumefaction evolving for a few years. The examination revealed cerebriform transverse occipital skin folds. The remaining physical examination was otherwise unremarkable. Given her chronic metabolic pathologies, secondary CVG was diagnosed.

Discussion

CVG is a rare benign condition first described by Alibert in 1837 characterized by proliferation and hypertrophy of the skin of the scalp or forehead. The affected skin then exhibits folds and furrows resembling the cerebral cortex convolutions. Unna introduced the term cutis verticis gyrata in 1907 [2].

This entity was afterward divided into different subtypes: a secondary and a primary form, then the primary form was subdivided into primary essential and non-essential. The primary essential form is the rarest form, characterized by an isolated cutaneous involvement with no associated pathology. It usually begins in adolescence and is more common in men than in women. Typically, as seen in case 1, primary CVG presents as symmetric scalp folds which usually extend anteroposteriorly from the vertex to the occiput and transversally in the occipital region. Terminal hair density is reduced on the folds, but not in the furrows. The primary non-essential form has been described in association with several neuropsychiatric pathologies like mental retardation, epilepsy, microcephaly, schizophrenia, encephalopathy, and other neurological malformations in addition to ophthalmic pathologies such as blindness, strabismus, and congenital cataract, the latter being the most frequent association. In primary non-essential CVG, the associated conditions do not have a clear pathophysiological link with the CVG.

Secondary CVG on the other hand is associated with conditions that may be implicated in modifications of the trophicity of the skin. It is slightly more common than the primary form. It can occur at any age without gender predominance. It is associated with a variety of underlying disorders. It may be caused by

general diseases comprise as liver affections, paraneoplastic syndromes, and pachydermoperiostosis or also associated with inflammatory dermatoses most commonly psoriasis and eczema [3]. Furthermore, various associated genetic conditions have been described such as neurofibromatosis, Noonan, Turner, Klinefelter and Ehlers–Danlos syndrome. It may also occur in association with endocrine etiologies, as seen in case 3, such as diabetes mellitus, myxedema, and the most described in the literature is acromegaly [3]. Among iatrogenic causes, treatment with minoxidil or misuses of anabolic substances were reported [4,5].

The pathophysiology of CVG remains unclear. On a structural level, it is suggested that the folds are caused by connective tissue septa formation between the skin and the galea. These septa prevent the expansion of the skin which bulges into folds [6]. In the primary form, an endocrine origin has been postulated as the disease usually manifests in postpubertal males. Cases reported in the literature are sporadic although some familial cases are described suggesting possible genetic factors in primary CVG. The physiopathology in the secondary form remains also unclear especially in general and inflammatory diseases. However, in the physiopathology of endocrine etiology, trophic hormones such as growth hormone (GH) and insulin growth factor 1 (IGF 1) were incriminated to be responsible for skin modifications with collagen thickening and hypertrophy of sebaceous glands.

When performed, histopathology of primary CVG shows normal skin. Thickened connective tissue with hypertrophy or hyperplasia of adnexal structures has also been described. Secondary CVG has variable histology representing the underlying disease. No treatment has proven its effectiveness in this entity. Abstention is the rule in the primary form. In all cases, regular rigorous hygiene of the skin folds and furrows is recommended to avoid maceration and infection. If treatment is requested by the patient because of an anesthetic complaint, medical and/or surgical treatment may be suggested. Surgical treatment seems to be the best option with a regularly reported efficacy. The undermining and relaxation incisions alter the relationship between the skin and the deeper structures and so result in the disappearance of the folds and furrows [7]. Medical treatment includes isotretinoin or systemic steroids. No treatment can prevent a recurrence.

In summary, CVG is a rare disease with mainly anesthetic impact. A rigorous clinical examination with a minimal workup is however required to eliminate a secondary or a primary non-essential form. If aesthetic prejudice is important, surgical treatment can be suggested.

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Drs Amal Chamli and Meriem Jones contributed to the first draft of the manuscript. Drs Amal Chamli, Meriem Jones,Takwa Bacha, and Noureddine Litaiem contributed to the literature search, analysis, and interpretation of the data. Dr Faten Zeglaoui critically revised the manuscript and gave final approval. All authors read and approved the final manuscript and agree to be finally accountable for ensuring the integrity of and accuracy of the work.

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Figure legends:

Figure 1: multiple symmetric skin folds extending from the vertex to the occipital area in a 28-year-old male

Figure 2: Cerebriform folds on the forehead of a 32-year-old male.