Cutis verticis gyrata and acromegaly

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Introduction

Cutis verticis gyrata (CVG) is a benign skin lesion which is also called "bulldog scalp or corrugated scalp". It is a very rare, innocuous, but anesthetic dermatologic condition where the scalp is folded and thickened in a way that resembles the brain surface [1,2]. The progressive and excessive development of the skin affects especially the scalp and/or the forehead [2,3] leading to profound folds and furrows which can be seat of fungal lesions. CVG is broadly classified in 2 forms: the primary and idiopathic CVG, and the secondary one which can be induced by some diseases [1,4] as in the following observation.

Case report

A 47-year-old man with a family history of metabolic syndrome was hospitalized for a pituitary tumor secreting growth hormone (GH) and uncommon skin lesions that began simultaneously six years ago. GH excess was already complicated by high blood pressure and diabetes mellitus (delete type 2). Our exploration discovered sleep apnea, and cardiomyopathy. The skin surface showed unusual lesions of the forehead and the anterior scalp: there were very thick horizontal and vertical folds separated by significant depressions with cerebral appearance (figure 1). Depressions or grooves are the seat of fungal lesions, but there was not any nevus and any lesion suspect of malignancy. Skin biopsy was refused by the patient.

Growth hormone concentration was high = 40 mIU/mL (N < 3), insulin like growth factor 1 (IGF1) = 1279 ng/mL (N = 175–375), but prolactin and the rest of pituitary function were normal except for gonadal deficit.

Cerebral MRI showed a pituitary tumor measuring 23 × 17 × 20 mm (figure 2). For treatment and follow-up, he received antifungal drugs plus anti-diabetic and anti-hypertensive regimens. Dopamine analogs were tried but in vain. Somatostatin's analogues: 120 mg/28 days for 6 months were inefficient. He was operated on three times, but iterative surgery failed to cure hypersomatotropism. Immunostaining confirmed the pure somatotroph secretion. He was proposed for radiotherapy, but he refused it, and then was lost in sight.

Discussion

Cutis verticis gyrata (CVG) was reported for the first time by Alibert in 1837. In 1843, Robert gave the definition and the entire description of this entity. Then after the word CVG was used by Unna in 1907 [1,5].

CVG diagnosis is based on clinical examination. Radiological images are not fundamental for the diagnosis, but when MRI is performed, it shows a characteristic "saw teeth" aspect of the scalp.

Figure 1

Acromegaly hand and skin lesions that look like the surface of the brain with transversal folds on the forehead and vertical ones on the scalp. Remark the deep grooves seat of fungal infection (not visible on the photos)

Funding: this report did not receive any specific grant from any funding agency in the public or commercial sector.
For etiologies, Polan and Butterworth who analyzed 195 cases in 1953 classified the skin lesions in two forms: primary or idiopathic CVG and secondary CVG. They noted that 47% were idiopathic and the rest were due and/or were associated to some diseases:
- the primary CVG (PCVG) is relatively rare. It is considered as a genetic disorder although genetic abnormalities are not defined yet. This form can be diagnosed in the womb [6] and is subdivided in two groups: essential and non-essential CVG:
  - the essential CVG or idiopathic CVG is generally not associated to other abnormalities. It is observed especially in men: 5–6/1 woman [4]. It usually occurs around the pubertal period, before 30 years old. The skin surface shows symmetrical grooves and histology demonstrates an increase in the connective tissue and skin appendages. But, sometimes skin biopsy did not show any abnormal structure [1],
  - the non-essential CVG is generally associated to some anomalies such as psychiatric or ophthalmological diseases [4]. Neuropsychiatric disorders observed in people with essential CVG are epilepsy, microcephaly, encephalopathy, and mental delay [1]. Ophthalmological abnormalities are numerous, but the most common is cataract;
- the secondary CVG (SCVG) affects all ages and both sexes. Several diseases can be responsible for this entity. They are separated in non-endocrine or general diseases, and endocrine diseases:
  - general diseases are nevi [7], fibroid tumors, obstructive pulmonary diseases, congenital heart diseases, liver affection, chronic inflammatory processes [8], and paraneoplastic syndromes. Anabolic substances abuse [5] and chemotherapy and/or radiotherapy can induce SCVG too [9]. HIV-related lipodystrophies, the syndrome of immunoglobulin excess, tuberous sclerosis, amyloidosis, acanthosis nigricans, trauma, Ehlers Danlos syndrome, syphilis and pachydermoperiostosis can be associated to SCVG too. In these diseases, the skin grooves are generally asymmetrical and histological examination shows subcutaneous hyperplasia of adipose tissue and sebaceous glands,
  - among endocrine diseases, hypersomatotropism condition is the most incriminated [10,11] as GH and IGF1 are both trophic hormones. GH and/or IGF1 are responsible for skin modifications with collagen thickening and hypertrophy of the sebaceous structures [9]. But, till now, no one knows why these folds and grooves are located only in the scalp and/or the face, and why is CVC absent in most people with pituitary gigantism and/or acromegaly. In our patient, the concomitant appearance of acromegaly and CVG pleads for GH excess etiology.
Other endocrine diseases, which are described in association with CVG, are diabetes mellitus, insulin-resistance, Noonan [4], Turner, and Klinefelter’s syndromes. Myxedema and empty sella may be observed with CVG too.
CVG medical treatment is based on iso-retinoids with or without corticoids, but their efficacy is not proved yet. A good and regular hygiene of the skin folds and depressions seems more fundamental for the prevention of microbial and fungal infections. Some medical shampoos can be very useful especially for the scalp.
Surgery remains the best treatment for the primary advanced disease [3,12–14]. Several methods have been proposed, but no one prevents recurrences. Some authors propose a combination of medical treatment with surgery, while others prefer wait and see attitude, especially for mild or moderate lesions.
For secondary forms, the ideal treatment should be elimination of the causative disease. For acromegaly, it seems that CVG noted in 14 to 30%, and slowly evolving stabilizes after surgical...
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resection of the pituitary adenoma or after a good control of GH/IGF1 excess by somatostatin’s analogues, dopamine agonists and/or GH receptors’ antagonists. This could not be demonstrated in our patient as surgery and medical treatment failed to cure hypopersomatotropism.

Plastic surgery has an important place in young patients with evolving CVG because of an important psychological prejudice. Concerning prognosis, CVG is a benign condition, except in rapidly evolving lesions or when there are nevi inside the grooves. Because, some nevi may become malignant. For this, a long-term follow-up is mandatory [15].

Acknowledgements: Authors would like to thank the patient who accepted publishing his photos and medical history, Pr. Boudjemaa Mansouri for providing cerebral MRI pictures, Amel Selimi and Nacer Tis for their technical assistance.

Disclosure of interest: the authors declare that they have no conflicts of interest concerning this article.

References

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Received 1st October 2014
Accepted 25 March 2015
Available online:

http://dx.doi.org/10.1016/j.lpm.2015.03.019
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