Cutis Verticis Gyrata – A Case Report

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ABSTRACT Cutis verticis gyrata (CVG) is a skin condition characterized by excessive growth of the skin of the scalp, resulting in furrows and folds which are similar to the gyri of the brain cortex. CVG can be classified into two forms: primary (essential and non-essential) and secondary. We report the case of a 53-year-old male patient with the primary type of CVG. The patient did not present with any cutaneous complication. His main complaint was the unaesthetic aspect of his scalp related to a psychological inferiority complex.

KEY WORDS: cutis verticis gyrata, hypertrophy, rare diseases, scalp dermatoses

INTRODUCTION

Cutis verticis gyrata (CVG) is a skin condition associated with excessive growth of scalp tissue, resulting in furrows and folds which are similar to the gyri of the brain cortex (1). Although Alibert first mentioned it in 1837, Robert described the condition in 1843. In 1907 Unna introduced the term "cutis verticis gyrata", which is still used today (2).

CVG can be classified into primary and secondary forms. The primary essential form is not associated with any other disorder except for the cutaneous alterations, whereas the primary non-essential type is associated with neurological and ophthalmological disorders (1,3). The secondary form occurs as a result of a local inflammatory or neoplastic process of the scalp or a systemic illness that produces pathologic changes in the scalp structure such as folliculitis, acne conglobata, or Darier's disease (4-6). The secondary form of CVG can also be caused by an underlying condition, such as amyloidosis, syphilis, acromegaly, pseudoacromegaly, myxedema, pachydermoperiostosis, neurofibroma, giant congenital melanocytic nevus (GCMN), or cerebriform intradermal nevus (3,7-10).

The male to female ratio is 5-6 to 1. The presentation, in the majority of cases, occurs before puberty, and only 10% of cases develop after 30 years of age. Secondary cutis verticis gyrata can appear at any age, depending on the underlying pathology. Some secondary forms, such as cerebriform intradermal nevus, may be present at birth (11).

The lesion is mostly asymptomatic, and 2-20 folds can occur symmetrically. The deep lines in the scalp typically present an anteroposterior disposition but may also present a horizontal disposition. CVG typically affects the vertex and occipital regions, but some forms can involve the entire scalp. The folds are soft and spongy and cannot be corrected by pressure or traction. The folds are usually symmetric in primary cutis verticis gyrata and asymmetric in secondary cutis verticis gyrata (11-13).

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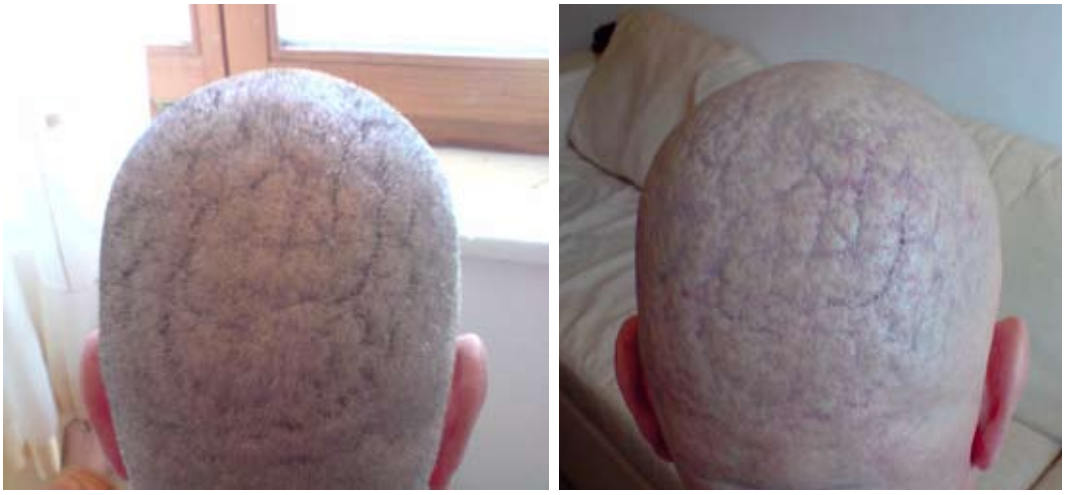

Figure 1. Cutis verticis gyrata. Scalp hypertrophy with anteroposterior and horizontal folds in the parietooccipital region.

CVG can be diagnosed by observing the characteristic morphological aspects of the patient´s scalp. However, in order to determine whether there are causative or associated diseases, the patient medical and family history are needed in addition to laboratory tests, radiological examination, and histological evaluation (12). A low free testosterone level was observed in some patients with primary CVG (14). In secondary CVG, laboratory tests depend on the associated disease. Imaging studies, such as magnetic resonance imaging (MRI) or computed tomography (CT), should be performed to determine or exclude any structural brain abnormalities (15). Histological findings are normal in most cases of primary CVG. In some cases, dermal collagen thickening, hypertrophy of the sebaceous structures, and multiple sweat glands and ducts may be present. In secondary CVG, histological findings depend on the underlying disease (2).

The treatment consists of local hygiene and surgical resection of skin excesses (1).

CASE REPORT

We report the case of a 53-year-old male patient who was referred to our Department because of folds in the scalp that had begun to develop 3-4 years earlier. The folds had gradually increased in size and were followed by erythematous pustules and scabs. The patient was treated with local and systemic antibiotics with no improvement in his condition. He had no family history of similar lesions and denied any previous history of inflammation of the scalp or signs and symptoms of neurologic or psychiatric disorders.

Physical examination revealed multiple soft and spongy folds localized on the vertex and occipital region of the scalp. Most of the folds presented an anteroposterior disposition. Folds were symmetric and did not extend into the nuchal region (Figure 1, Figure 2). No areas of alopecia were observed, nor were there differences in hair distribution between the affected areas and areas of normal skin. The rest of the physical examination did not reveal any other significant skin lesions.

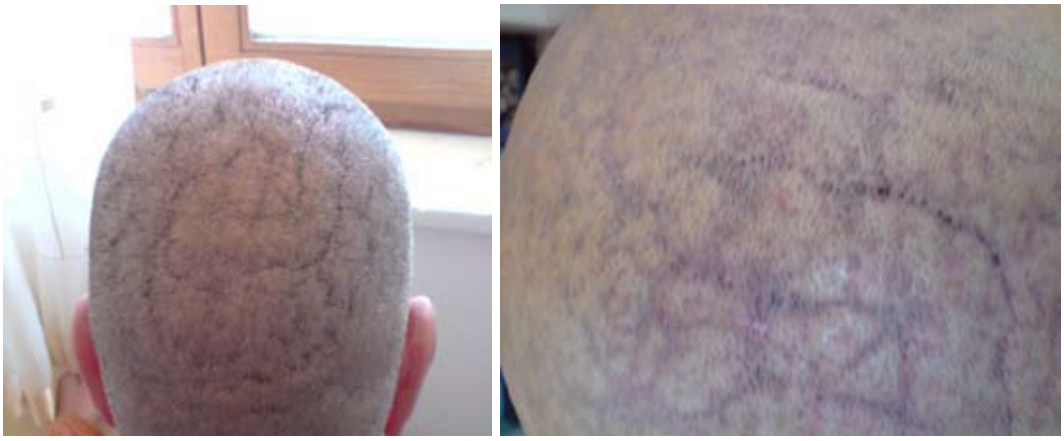

Figure 2. Cutis verticis gyrata. Horizontal disposition in the occipital region.

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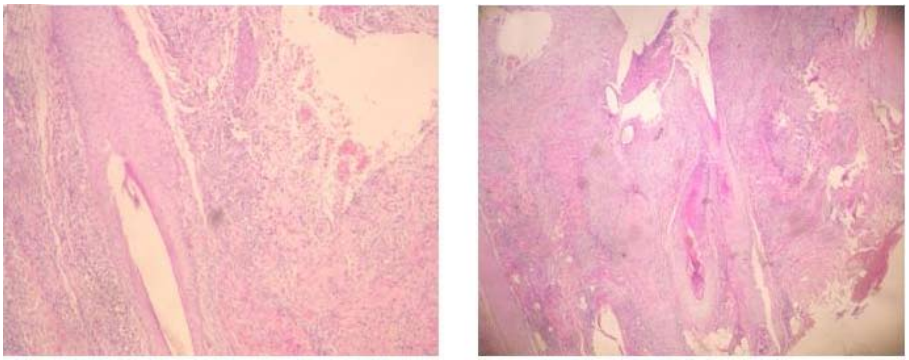

Figure 3. Histopathological skin sample showing central flask-shaped epidermal ulceration covered with crust and a fibrin rich ulcer bed, with associated vascular proliferation (hematoxylin and eosin, ×100).

A full laboratory workup was performed. The complete blood count showed an elevated level of red blood cells count 5.46×10^{12}/L (normal values 3.8-5.08×10^{12}/L). Biochemistry laboratory tests showed increased levels of alanine amino transferase (ALT), cholesterol, triglycerides, and *low-density lipoprotein (*LDL) cholesterol. Total immunoglobulin E (IgE), eosinophil cationic protein (ECP), urinalysis, computed tomography (CT), mycological scalp culture, skin scrapings, and ophthalmologic exams revealed no significant alterations.

Microbiological examination showed increased level of serum antistreptolysin (AST-O) 1600 IU (normal levels <200 IU), while the serum Waaler-Rose test and *Treponema pallidum* hemagglutination test (*TPHA*) revealed no significant alterations.

A skin biopsy specimen showed no signs of underlying disease (Figure 3).

Based on the clinical features and the results of the tests performed, we established a diagnosis of primary essential cutis verticis gyrata.

DISCUSSION

CVG is a term that refers to a pattern of redundant skin on the scalp that exhibits deep furrows and convolutions. The hypertrophy and folding of the skin produces a gyrate appearance (12).

In this case report we reported a case of a 53-year old male patient with primary essential CVG. The patient started developing multiple scalp folds 3-4 years prior to referral, with gradual increase in size. Primary CVG occurs predominantly in men and typically after puberty. Most primary cases develop before the age of 30. In our case, the condition started developing after the age of 30 (11).

Our patient had multiple scalp folds localized on the vertex and occipital regions. Most of the folds presented an anteroposterior disposition, except for the folds in the occipital region which presented a horizontal disposition. Folds were symmetric, soft, and spongy. The literature states that CVG typically affects the vertex and occipital regions. In primary CVG, folds are usually symmetric (2). Based on the clinical features and the results of the tests performed, we established a diagnosis of primary essential cutis verticis gyrata.

The differential diagnosis of primary essential CVG includes the nonessential form, pachydermoperiostosis, acromegaly, intradermal nevus, and other less frequent causes of CVG (2). Primary nonessential CVG is associated with psychiatric, neurological, and ophthalmological disorders. Studies showed a high prevalence of primary CVG in men, which has often been associated with mental retardation (13). Pachydermoperiostosis (PDP) is a rare genodermatosis, characterized by pachydermia, digital clubbing, and periostosis. It has often been confused with primary CVG, but PDP affects not only the skin of the scalp but also the skin of the face, hands, and feet and is accompanied with periosteal features. Overgrowth of the scalp manifesting as CVG is observed in 10-15% of patients with acromegaly (17,18). Intradermal nevus is a disorder usually present at birth or early in life and is a rare cause of secondary CVG or pseudo-CVG. Cerebriform intradermal nevus is an asymmetric, skin colored or slightly pigmented tumor, usually localized in the parietal or occipital areas of the scalp (3).

The treatment for primary essential CVG may be symptomatic or surgical. Education regarding local hygiene care is necessary to avoid secretion accumulation resulting in an unpleasant odor. Surgical treatment may be performed with the goal of improving the clinical aspect, since quality of life might be compromised due to an unesthetic aspect (19-21).

The patient was informed of the benign nature of the lesion, and no treatment was administered.

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