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# Cutis verticis gyrata in a patient with multiple basal cell carcinomas; case presentation and review of the literature

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## Abstract

Cutis verticis gyrata is a rare disease characterized by convoluted folds and deep furrows of the scalp, resembling the gyri and sulci of the cerebral cortex. Basal cell carcinoma is the most frequent cancer in Caucasians, patients frequently presenting multiple tumors. We report the case of a 62 year old male, Caucasian patient, from the urban area, who addressed the dermatology department of our hospital for multiple tumors located on the face and upper trunk. A careful examination revealed cerebriform folding of the skin of the scalp. Neurological, psychological, ophthalmological and endocrine disorders were disproven. The patient was diagnosed with cutis verticis gyrata based on the clinical picture and anamnesis, and basal cell carcinoma based on the histopathological examination. Since cutis verticis gyrata predated the BCCs by four decades, and no other conditions were associated, the patient was diagnosed with primary essential cutis verticis gyrata.

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**Keywords:** cutis verticis gyrata, basal cell carcinoma, neurologic abnormalities



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## **Introduction**

Cutis verticis gyrata is a rare disease characterized by convoluted folds and deep furrows of the scalp, resembling the gyri and sulci of the cerebral cortex. It is classified into primary cutis verticis gyrata and secondary cutis verticis gyrata. The primary form is also divided into an essential form, which associates no other disorders, and a non-essential form, which associates neurologic and/or ophthalmologic abnormalities. It is more frequent in males. Basal cell carcinoma (BCC) is a malignant tumor which arises from the basal layer of the epidermis and the pilosebaceous follicle. It is a locally invasive tumor which only rarely metastasizes. It is the most frequent cancer in Caucasians. Patients often have multiple primary BCCs (1, 2).

## **Case presentation**

We report the case of a 62 year old male, Caucasian patient, from the urban area, who addressed the dermatology department of our hospital for multiple tumors located on the face and upper trunk. The lesions had appeared approximately four months beforehand. The personal history revealed that the patient had been diagnosed with multiple basal cell carcinomas and metatypical carcinomas several times before and the lesions had been surgically excised or electrocauterized. The patient admits to having

exposed himself to sun during childhood and adulthood. The physical examination revealed a healthy appearing patient, skin type II on Fitzpatrick scale, blue eyes and red hair. The blood pressure and heart rate were within normal range.



*Figure 1.* Multiple BCCs on the face and scars following previously excised tumors

The clinical examination revealed multiple tumors, of variable sizes, ranging from 5 mm to 1.2 cm in diameter. Some of the lesions appeared as erythematous nodules with visible telangiectasia while others were ulcerated and covered by hematic crusts. The lesions were distributed on the lower lid, nose, ear, cheeks and upper trunk. The patient also presented several atrophic scars following the previously excised tumors (*Fig. 1*). A careful examination of the patient also revealed cerebriform folding of the skin of the scalp (*Fig. 2*). Further inquiry revealed that the folding of the scalp had appeared over 40 years before during

adolescence, but was ignored by the patient because it was asymptomatic.

Laboratory findings were within normal range. The patient was sent for an endocrinology examination which ruled out any endocrine disorders, including acromegaly, myxedema and Graves's disease. Some of the tumors were excised, the bioptic specimen being sent for histopathological examination, while other lesions were electrocauterized. The histopathological examination confirmed the clinical diagnosis of basal cell carcinoma. The cerebriform folding of the skin was diagnosed as cutis verticis gyrata and required no treatment. The patient remains under our supervision for early detection of new BCCs.

## Discussions

*Cutis verticis gyrata* (CVG), also known as paquidermia verticis gyrata, cutis verticis plicata, cutis sulcata, cutis capita striata or "bulldog" scalp syndrome, is a rare condition characterized by ridges and furrows resembling the surface of the brain. It was first mentioned in the medical literature by Ailbert, in 1837, who called it "cutis sulcata". However, it was first clinically described by Robert in 1843. The term "cutis verticis gyrata" was introduced in 1907 by Unna (3- 7). Males are more frequently affected than females, the estimated prevalence in males being 1/ 100.000 and in females 0.026/ 100.000.



Figure. 2. Cutis verticis gyrata: cerebriform folding of the skin of the scalp

The prevalence seems to be higher in patients with neuropsychiatric disorders and primary essential CVG is very rare. A study performed by Åkesson between 1961 and 1962 in institutions for the mentally deficient in Sweden showed a prevalence in severely retarded men of 0.2%. Chen studied 1244 male patients hospitalized in a mental institute in Taiwan and reported a prevalence of primary CVG of 2% in patients with chronic schizophrenia. CVG usually occurs during late childhood or adolescence, generally before the age of 30, and slowly becomes more accentuated in time (2, 4, 5, 8-10).

Polan and Butterworth classified the disorder in 1953 into primary CVG and secondary CVG. In 1984 Garden et al further divided the primary form into an essential form, which associates no other conditions, and a non-essential form, associated

with neurologic and/ or ophthalmologic abnormalities (1, 11, 12).

The pathogenesis of the disorder is unknown. According to some authors, it might be an autosomal dominant condition determined by mutations of FGFR2, a gene located on chromosome 10q22 which encodes a transmembrane tyrosine kinase and can function as a mitogenic, angiogenic and inflammatory factor (13).

Primary essential CVG is very rare and can only be diagnosed after excluding any other conditions. Primary non-essential CVG is associated with neurologic abnormalities, such as mental retardation, seizures, cerebral palsy, or ophthalmologic abnormalities like cataracts, optic atrophy, strabismus or retinitis pigmentosa. Clinically, scalp folds are symmetric and have a longitudinal direction between the vertex and occiput. The hair is usually not affected. The histopathological examination usually shows no alterations but may present increased collagen bundles and hypertrophy of the adnexal structures (1, 2, 5).

Secondary CVG is less frequent than primary non-essential CVG and has a similar distribution between the two sexes (1). It has been associated with several disorders (*Table 1*) and the clinical aspect depends on the associated disorders (5). As compared to primary CVG, in secondary CVG the skin folds can have an asymmetrical

appearance. Also, the disease can occur at any age (14). Acromegaly and pachydermoperiostosis are two of the disorders most often associated with CVG (14).

Pachydermoperiostosis (Touraine-Solente-Golé syndrome) is a rare hereditary syndrome characterized by finger clubbing, pachydermia, wrinkling of facial features and, sometimes, cutis verticis gyrata. Patients often associate seborrhea, hyperhidrosis, acne and folliculitis. Facial and pubic hair is rare. It is sometimes confused with primary CVG. However, primary CVG only affects the skin of the scalp while pachydermoperiostosis also affects the face, hands and feet (15-17). Rosenthal-Kloepfer syndrome (acromegaloid phenotype with cutis verticis gyrata and corneal leukoma) is a rare disorder with autosomal dominant inheritance. Patients have acromegaloid facial features (tall, with large hands, large feet and chin) but no endocrine abnormalities are detected. The sella turcica is normal in size. The scalp skin undulation has a sagittal orientation (18). In the case we are presenting, the disorder was asymptomatic and had first been noticed by the patient during adolescence. Clinically, it manifested as folding of the scalp with a longitudinal direction between the vertex and occiput. The patient did not associate any neurological or ophthalmological disorders. The endocrinology consult also ruled out any endocrine disorders. The patient did, however present several basal cell carcinomas.

**Table 1.** Secondary cutis veritics gyrata associations

Endocrine disorders	Systemic disorders	Hereditary disorders
<ul style="list-style-type: none"> <li>➤ Acromegaly</li> <li>➤ Pseudoacromegaly</li> <li>➤ Myxedema</li> <li>➤ Grave's disease</li> <li>➤ Diabetes mellitus</li> </ul>	<ul style="list-style-type: none"> <li>➤ Syphilis</li> <li>➤ Leukemia</li> <li>➤ Neurofibromatosis</li> <li>➤ Acanthosis nigricans</li> </ul>	<ul style="list-style-type: none"> <li>➤ Turner syndrome</li> <li>➤ Noonan syndrome</li> <li>➤ Ehler-Danlos syndrome</li> <li>➤ Klinefelter syndrome</li> </ul>
<b>Inflammatory diseases of the scalp</b>	<b>Benign tumors</b>	➤ Fragile X syndrome
<ul style="list-style-type: none"> <li>➤ Eczema</li> <li>➤ Psoriasis</li> <li>➤ Impetigo</li> <li>➤ Erysipelas</li> <li>➤ Pemphigus</li> <li>➤ Darier's disease</li> <li>➤ Folliculitis</li> <li>➤ Impetigo</li> <li>➤ Acne conglobata</li> </ul>	<ul style="list-style-type: none"> <li>➤ Dermatofibroma</li> <li>➤ Neurinoma</li> <li>➤ Cerebriform intradermal nevus</li> <li>➤ Hamartomas</li> </ul>	<ul style="list-style-type: none"> <li>➤ Pachydermoperiostosis</li> <li>➤ (Touraine-Solente-Golé syndrome)</li> <li>➤ Rosenthal-Kloepfer syndrome</li> <li>➤ Tuberous sclerosis</li> <li>➤ Beare Stevenson Syndrome</li> </ul>
	<b>Malignant tumors</b>	
	<ul style="list-style-type: none"> <li>➤ Malignant melanoma</li> <li>➤ Angiosarcoma</li> <li>➤ Fallopian tube carcinoma</li> <li>➤ Infiltrating ductal carcinoma</li> </ul>	

*Basal cell carcinoma* is the most common cancer from both natural and artificial light sources. in Caucasian patients. It is a malignant tumor which arises from the basal layer of the epidermis and the pilosebaceous follicle. The mortality associated with BCC is very low because this tumor only very rarely metastasizes. However, it is locally invasive and, since it is often located on the face, the morbidity is high. The incidence of BCC is increasing worldwide (19- 21).

The most important risk factor for basal cell carcinoma is UV exposure, especially UVB, fair skin, Fitzpatrick phototype I and II, blond or red hair and blue eyes. Some genetic syndromes

presenting multiple BCCs have also been described, including Gorlin syndrome, Bazex syndrome and Rombo syndrome (1, 18, 21).

Our patient presented multiple tumors, of variable sizes, ranging from 5 mm to 1.2 cm in diameter. Some of the lesions appeared as erythematous nodules with visible telangiectasia while others were ulcerated and covered by hematic crusts and were distributed on the lower lid, nose, ear, cheeks and upper trunk. He had several risk factors for developing BCCs: fair skin type, red hair and blue eyes. Also, he admits having exposed himself to sun during childhood and adulthood. The high number of BCCs is particular. Therefore, the differential diagnosis included genetic syndromes associated with BCCs. However, apart from the high number of BCCs, the patient presented no criteria for the diagnosis of any genetic disorder.

CVG does not generally require any treatment. However, depending on the symptomatology, several therapeutic methods have been tried with variable results. Corticosteroids, antihistamines, radiotherapy, sleep therapy or psychotherapy failed to prove their efficacy, therefore surgical excision remaining the treatment of choice. The tissue expansion method can be tried before surgical excision. This procedure requires the implantation of an expander under the skin and saline injections. After a few months, the lesions are excised and the resulted flaps are used

to repair de defect. In small lesions, surgical excision can be performed per primam. Subcutaneous incision can also be tried in patients with primary CVG (5, 22).

Several treatment methods are available for BCCs. Surgical treatment is the treatment of choice and is associated with the lowest recurrence rate. Mohs micrographic surgery is recommended when tissue conservation is very important. Electrodesiccation and curettage can also be performed in small lesions with good cosmetic results. Other treatment methods include cryosurgery, imiquimod, 5-fluorouracil, photodynamic therapy and radiation therapy. Vismodegib, an inhibitor of the smoothed receptor in the hedgehog pathway, is a novel treatment reserved for metastatic and locally advanced BCCs (20, 23, 24).

In the case we are presenting, the larger BCCs were surgically excised and the smaller ones were electrodesiccated. CVG was asymptomatic and did not require any treatment. The patient remains under our supervision for the early detection of any new BCCs.

## **Conclusions**

Cutis verticis gyrata is a rare condition. The primary essential form is the most infrequent, with only a few cases being described in the medical literature. The secondary form is more prevalent and can be combined with several disorders. In the

case we are presenting, all the disorders frequently associated with CVG were disproven, as were all the rare ones. The patient did associate however several BCCs. BCC has never been reported in the medical literature in association with CVG and we do not believe that due to their presence the patient should be diagnosed with secondary CVG. BCC is a very frequent tumor and our patient had several risk factors for developing this disorder. Also, it first occurred decades after CVG and did not modify in any way the clinical aspect or symptomatology. Therefore, we report a very rare case of primary essential cutis verticis gyrata, a coincidental finding in a patient with multiple BCCs.

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