Case report

Cutis verticis gyrata resection with scalp reconstruction: a case report

Ressecção de cutis verticis gyrata com reconstrução de couro cabeludo: relato de caso

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■ ABSTRACT

Cutis verticis gyrata is a disease characterized by hypertrophy of the skin on the scalp, leading to the formation of folds and sacculations that resemble the gyrus of the cerebral cortex. It most commonly affects males and develops after puberty. It can occur alone or in association with various underlying conditions and treatments, including neuropsychiatric disorders, eye abnormalities, or inflammatory conditions. The disease management can range from conservative conduct with correct asepsis of the areas of folds and surgery if requested for psychological or aesthetic reasons. The present study aims to report the case of an adult patient with cutis verticis gyrata who underwent surgical treatment for complete resection of the lesion, followed by reconstruction with skin flaps and serial grafting, together with a vacuum negative pressure dressing.

Keywords: Adult; Congenital abnormalities; Scalp; Hypertrophy; Aesthetics; Plastic surgery; Allografts; Vacuum.

■ RESUMO

A *cutis verticis gyrata* é uma moléstia que se caracteriza pela hipertrofia da pele do couro cabeludo, levando à formação de dobras e saculações que se assemelham aos giros do córtex cerebral. Acomete mais comumente o sexo masculino e se desenvolve após a puberdade. Pode ocorrer isoladamente ou em associação com uma variedade de condições e tratamentos subjacentes, incluindo distúrbios neuropsiquiátricos, anormalidades oculares ou condições inflamatórias. O manejo da doença pode incluir desde conduta conservadora com a assepsia correta das áreas de dobras bem como cirurgia, se solicitado por razões psicológicas ou estéticas. O presente estudo tem por objetivo relatar o caso de um paciente adulto com *cutis verticis gyrata* submetido a tratamento cirúrgico para ressecção completa da lesão, seguida de reconstrução com retalhos cutâneos e realização de enxertias seriadas, juntamente com curativo por pressão negativa a vácuo.

Descritores: Adulto; Anormalidades congênitas; Couro cabeludo; Hipertrofia; Estética; Cirurgia plástica; Aloenxertos; Vácuo.

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Article received: August 12, 2020.
Article accepted: April 23, 2021.

Conflicts of interest: none.

DOI: 10.5935/2177-1235.2021RBCP0134

INTRODUCTION

CVG is a disease characterized by the excessive growth and thickening of the skin on the scalp, causing the formation of sulci that resemble the turns of the cerebral cortex[1]. In addition, affected individuals demonstrate visible folds, furrows or creases on the surface of the upper part of the scalp, which can vary in quantity from 2 to 10. A typically spongy/soft consistency usually affects the parietal and occipital regions of the skull, but in some cases rarer, it can affect the entire scalp.

Jean-Louis-Marc Alibert first reported the condition in 1837, who called it cutis sulcata. In 1907, it was named by Paul Gerson Unna, who gave it other names such as Robert-Unna syndrome, bulldog scalp, corrugated skin, cutis verticis plicata and pachydermia verticis gyrata. Although its etiology is still unknown, what is known is that the condition is not exclusively congenital. There are two classifications of CVG: primary and secondary[2,3].

The primary form can be subdivided into essential and non-essential. The essential primary form is not

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associated with neurological and/or ophthalmological alterations, only the excessive formation of skin folds on the scalp[3,4]; it appears at puberty and exclusively in men; the type of inheritance is uncertain. Non-essential CVG corresponds to 0.5% of patients with cognitive impairment; cerebral palsy, epilepsy, cataract and amaurosis may be present[2].

The secondary forms of CVG are most frequently represented by dermopathies, such as pachydermoperiostosis, acromegaly, intradermal cerebriform nevus, etc. However, it can also be due to a range of disorders or twhe exacerbated use of certain drugs that mimic acromegaly, melanocytic nevus and inflammatory processes.

Alopecia can occur over time as the scalp thickens, although hair in any furrow remains normal.

In children, cases of CVG, regardless of its etiology, are very rare, and the few reports are due to the non-essential primary form, association with genetic syndromes and familial cases. In the world of medical literature, there is no report of a child with essential primary CVG. Due to the (apparent) rarity of the disease, research remains limited.

CASE REPORT

AFDS male patient, 30 years old, born in Rio de Janeiro. He complained about the unsightly appearance of the shape of his skull due to the "folds" it presented, in addition to the constant need to wear a cap to alleviate the embarrassment he felt due to the eyes of others when walking through the streets. He informed that he had sought other plastic surgery services before, but he was not submitted to a surgical procedure for various reasons.

On physical examination, during palpation, it was observed that the right parietal/temporal and occipital region of the skull had furrows and skin hypertrophy covered by hair, respecting the lines of capillary implantation (Figure 1).

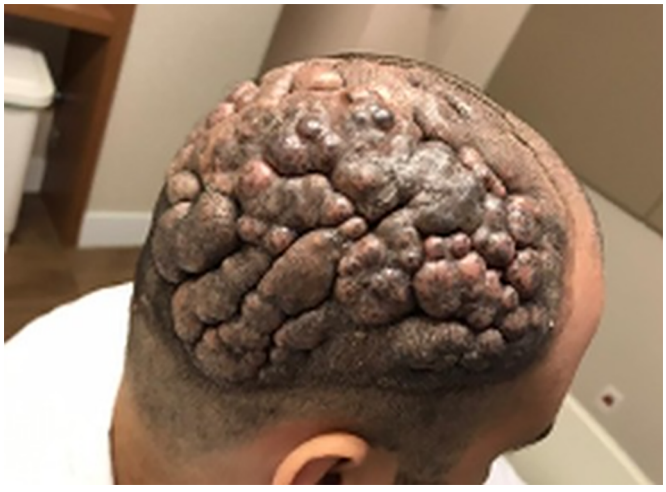


Figure 1. Pre-operative.

The patient did not complain of pain or compressive symptoms. The surgical procedure was performed under general intravenous anesthesia + sedation. Trichotomy of the entire head was performed, and subsequent demarcation of the area to be removed (Figure 2).

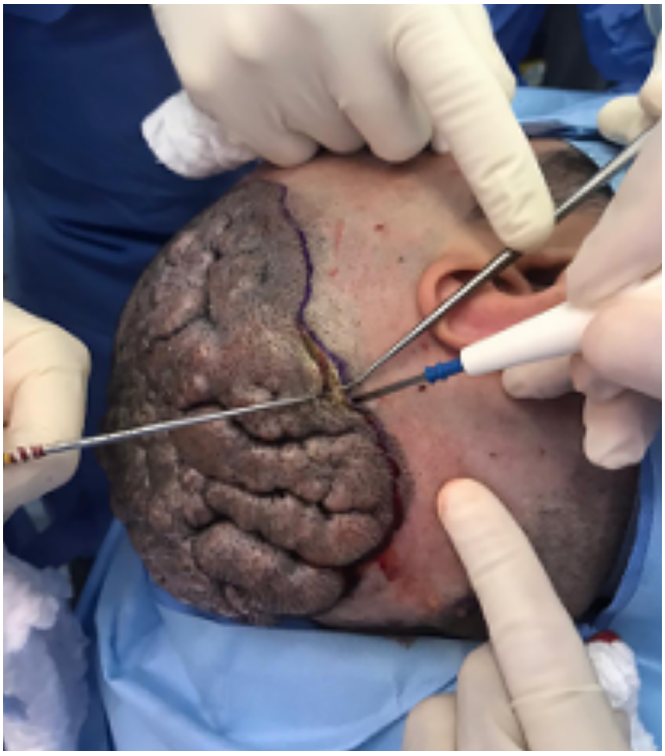


Figure 2. Marking and resection of the lesion.

A technique was used to resect the entire lesion with a safety margin of 0.5cm in thickness and depth. A lesion with very heterogeneous characteristics was observed during the incision, varying the degree of infiltration in the skullcap. In some regions of the parietal region, it was necessary to remove part of the periosteum.

Despite the difficulty of hemostasis, the lesion was excised entirely and in a single piece measuring 15x11cm, and the product of the resection was sent for anatomopathological analysis and CVG was confirmed (Figure 3).

After excision, a large open area with an irregular surface was observed. Then, a dressing with non-adherent petrolatum gauze was performed, using negative pressure therapy (@VAC), using a peristoma paste for better sealing (Figure 4).

The patient was surgically re-approached on the 3rd postoperative day, with hemostasis review and @ VAC replacement, in addition to skin traction with 3-0 nylon "U" stitches to approximate the wound edges (Figure 5).

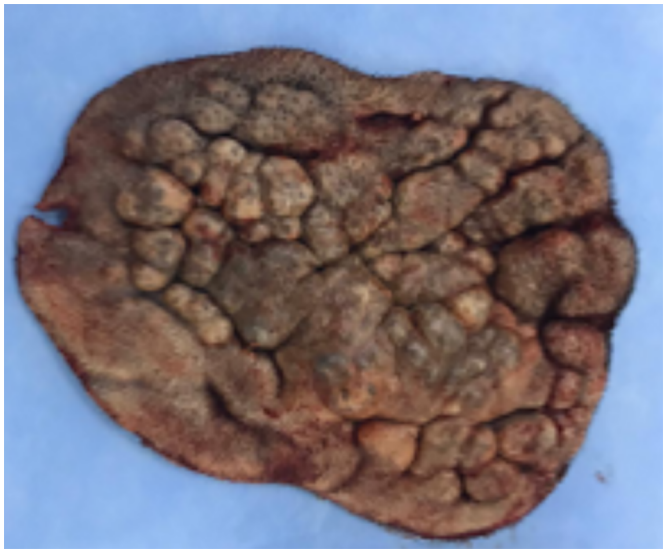
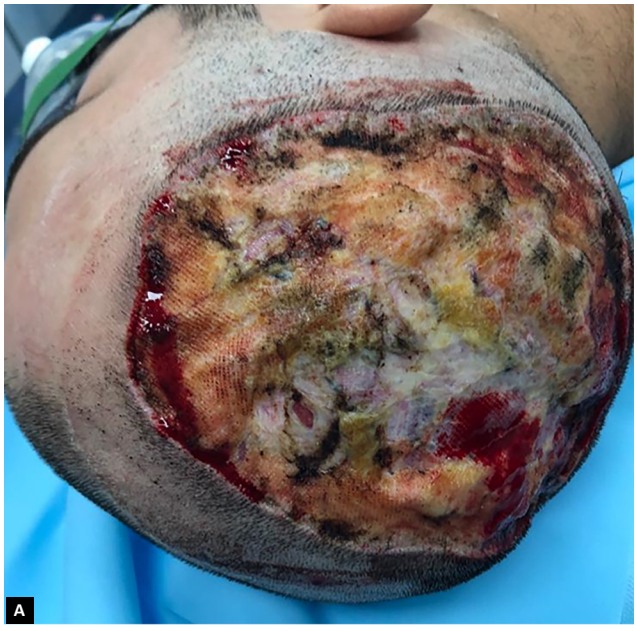


Figure 3. Excised part.



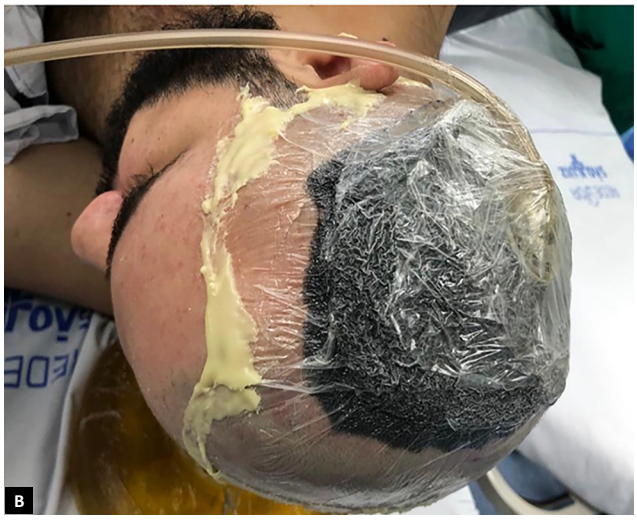


Figure 4. A. Open area in the right parietal-temporal region after lesion resection; **B.** Negative pressure dressing application.

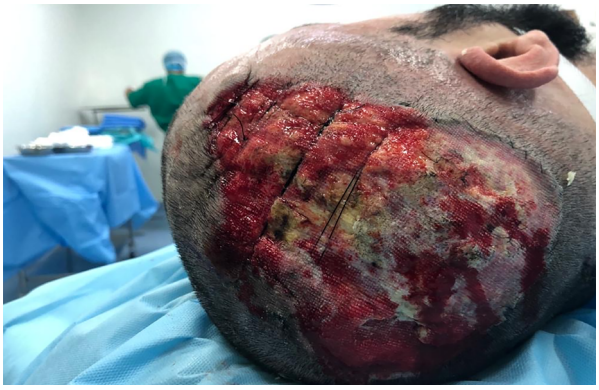


Figure 5. Cutaneous distraction technique for approximation of the wound edges.

This surgical schedule was followed for three weeks, changing the dressing under vacuum every 3-5 days until we had enough granulation tissue in the surgical wound. Following up, on the 21st postoperative day, the process of grafting the wound + production of skin flaps for advancement by rotation of the temporoparietal region began. The left parietal and temporal regions were chosen as donor areas. Using an electric dermatome, strips of skin with a partial thickness of 0.4mm were removed from these sites and fixed to the wound bed with 4-0 prolene thread with an atraumatic needle, ending with a vacuum dressing. (Figures 6A, B and C).

Another four surgeries were necessary to finish grafting the entire exposed surface after removing the lesion. Thus, after 11 surgical approaches in a total of 38 days of hospitalization, the therapeutic proposal was completed, and the patient was discharged for outpatient follow-up (Figures 7A, B and C - postoperative hospital discharge, 15 days and three months, respectively).

DISCUSSION

CVG is a rare entity of unknown etiology, often appearing after puberty, suggesting hormonal stimulation. It may have an autosomal recessive or dominant inheritance. It most often affects the vortex and occipital region, but it can affect the entire scalp. The true CVG can be idiopathic, associated with endocrine disorders (acromegaly, myxedema or cretinism), or syndromes such as pachydermoperiostosis, Beare-Stevenson syndrome or Turner syndrome[1].

In pseudo-CVG, we have the following differential diagnoses: cerebriform dermal nevus, focal mucinosis, neurofibroma and cutis leukemia. Optical microscopy provides a secure diagnosis. The only possible treatment is surgical correction.

Despite what is described in the medical literature, the patient did not present any phenotypic alteration or associated symptoms, possibly being a case of primary essential CVG. The pathological

Figure 6. A. Wound with granulation tissue; **B.** Donor area; **C:** Partial skin grafting and reconstruction with skin flaps performed

examination confirmed the clinical suspicion of CVG. The patient reported having the lesion since childhood and that the condition was progressive. Only in adulthood, for reasons solely and exclusively aesthetic, did he seek professional help, having difficulty finding a specialized medical service.

There are case reports in the medical literature where the treatment of CVG was performed in only two surgical times; where in the first surgery, tissue expanders are used in the skullcap. Previous skin preparation using expanders allows for more significant tissue gain for closure of the surgical wound after CVG excision. After six months of expansion (on average), it becomes possible to remove the tumor and close the open area with flap advancement techniques in a second surgical time[5].

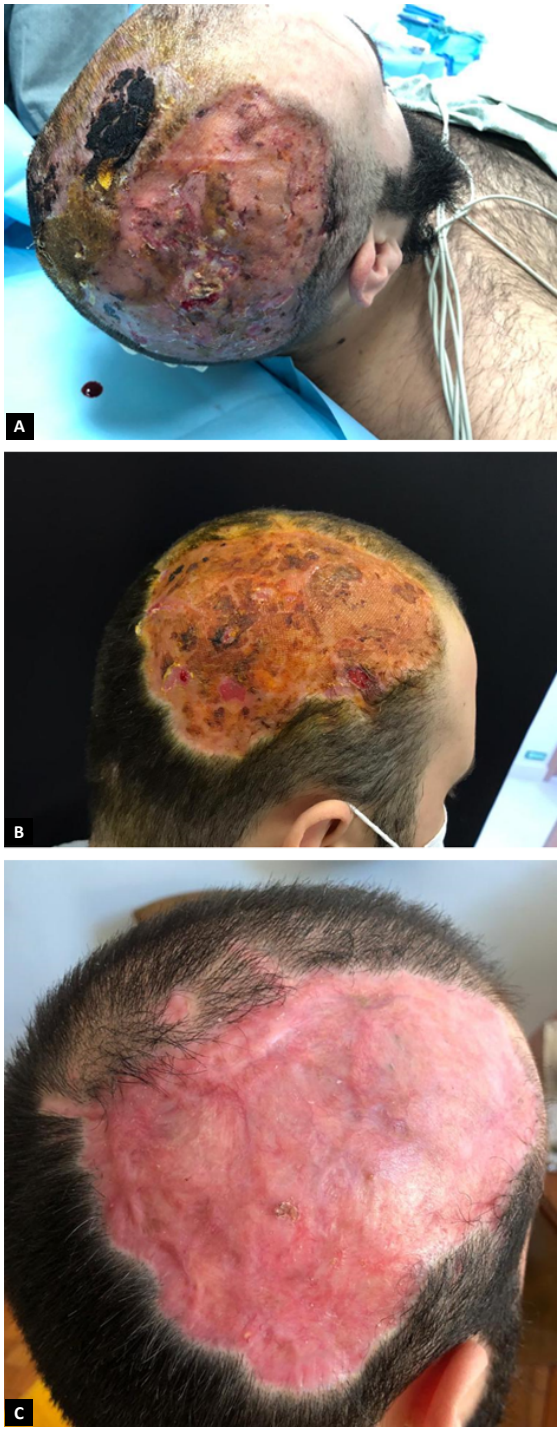


Figure 7. Postoperative: **A.** Hospital discharge; **B.** 15 days; **C.** 3 months.

This technique requires greater patient cooperation and psychological counseling, given that not everyone accepts the use of tissue expanders in the head region due to exposure. Thus, we chose not to use expanders at first, despite the more extended hospital stay and a greater number of surgeries, we believe that sequential grafts have a good aesthetic result with low rates of complications.

CONCLUSION

Cutis verticis gyrata is a rare entity, usually approached in two surgical stages using tissue expanders previously and followed by surgery to resection/close the lesion. However, this clinical case demonstrated that CVG could be treated more optimally with the removal of the tumor and the performance of sequenced skin grafts. Correct wound management combined with negative pressure vacuum therapy enabled an acceptable esthetic result. Despite the longer hospital stay and multiple surgical approaches, the patient was satisfied with his appearance, noting that his case was resolved within 38 days of treatment. It is also noteworthy that the patient is being followed up on an outpatient basis and undergoing psychological counseling to decide whether or not to accept the placement of tissue expanders for a better aesthetic result and to mitigate the alopecia area.

COLLABORATIONS

AMP Analysis and/or data interpretation, Conception and design study, Data Curation, Investigation, Methodology, Project Administration, Realization of operations and/or trials, Software, Visualization, Writing - Original Draft Preparation.

MOS Data Curation, Final manuscript approval, Project Administration, Realization of operations and/or trials, Resources, Supervision, Validation, Visualization, Writing - Review & Editing.

PRLC Data Curation, Investigation, Realization of operations and/or trials, Supervision.

BMV Data Curation, Investigation, Realization of operations and/or trials.

LSGC Data Curation, Investigation, Realization of operations and/or trials.

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