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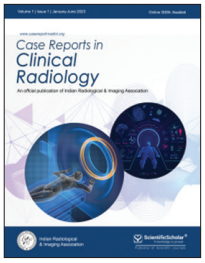

Case Report

Cerebriform intradermal nevus manifesting as cutis verticis gyrata

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ABSTRACT

Cutis verticis gyrata (CVG) is a rare scalp condition characterized by scalp skin redundancy that manifests as thickening of scalp skin with furrows and convoluted folds, giving out a cerebriform pattern of external appearance. CVG might manifest either as primary or secondary to an underlying cause. This case demonstrates the typical imaging findings of CVG that was provisionally diagnosed on imaging and confirmed on histopathology as secondary to cerebriform intradermal nevus in a healthy adolescent girl presenting with history of focal irregular scalp thickening since birth.

Keywords: Cutis verticis gyrata, Cerebriform intradermal nevus, Gyriform scalp lesion

INTRODUCTION

Cutis verticis gyrata (CVG) is a rare cutaneous disorder, characterized by cerebriform appearance of the scalp skin with convoluted folds and deep furrows.[1] It is usually located on the scalp, but other locations such as neck, lower limbs, buttocks, scrotum, or back have also been reported.[2,3]

CVG can be considered a manifestation of variety of causes rather than an individual disease. The worldwide prevalence of CVG is difficult to establish due to the extreme rarity of this condition. Of the known reports of CVG, there is a definite male predilection, with an estimated prevalence of 1 in 100,000 male population and 0.026 in 100,000 female population.[4] CVG can occur at any age and in either sex; however, there are several reports of primary essential CVG presenting in pre-pubertal men. Till date, there is no single established pathogenesis for this disorder; however, there are many systemic diseases which may be found in association with CVG. Accordingly, CVG is classified into primary and secondary form.

The objective of this paper is to describe a rare case of cerebriform intradermal nevus (CIN) presenting as CVG in a healthy adolescent girl, discussing its clinical and radiological findings.

CASE REPORT

A 13-year-old healthy adolescent girl with good scholastic performance presented with irregular thickening of the left anterior half of scalp since birth, the irregularly thickened area was initially around 3 × 3 cm and has gradually progressed to the present size spanning an approximate dimension of 10 × 8 cm [Figure 1]. Patient denies any other associated history of

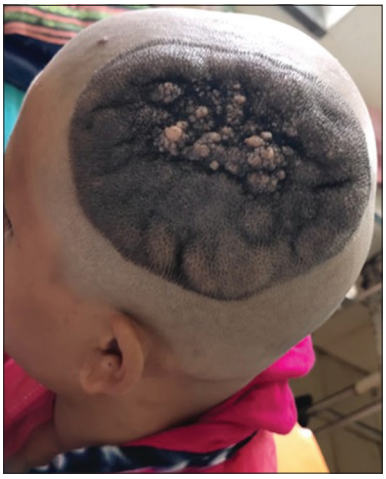

Figure 1: Focal irregular thickening of the scalp region giving a cerebriform pattern of appearance.

seizures, visual disturbances, neuropsychiatric conditions, scalp injuries, chronic dermatitis, usage of anabolic steroids or any kind of surgeries related to the scalp region and there was no history of similar lesions among family members.

On examination the irregularly thickened scalp skin had ridges and furrows giving a cerebriform pattern of appearance with few non-pigmented proliferative growths on the lesion. On ultrasound of the scalp region [Figure 2a] – the lesion was ill defined, hyperechoic and was occupying the subcutaneous plane of left scalp area and was not taking any vascularity on color Doppler [Figure 2b], thus ruling out any underlying vascular malformation.

Contrast-enhanced computed tomography [Figure 3] and magnetic resonance imaging of brain [Figure 4] revealed a focal area of extracranial, irregular, cerebriform pattern of thickening, involving dermal, epidermal, and adipose layers of scalp in left fronto-parieto-temporal regions and was showing subtle post contrast enhancement as compared to the surrounding scalp region with no obvious evidence of feeding arteries/draining veins noted with respect to the lesion. The lesion was not having any intracranial extension. Underlying bony cortex appeared intact with no features of cortical breach/calvarial thickening and the underlying brain parenchyma was normal.

In light of these findings a provisional radiological diagnosis of CVG, possibly secondary to CIN was made. The patient underwent complete excision of the scalp lesion and was covered with partial thickness skin graft, the excised specimen histopathologically [Figure 5] revealed nevus cells arranged in nests, cords and lying singly with few of them containing melanin along with entrapped adnexal structures in the entire dermal layer in its full thickness. A final

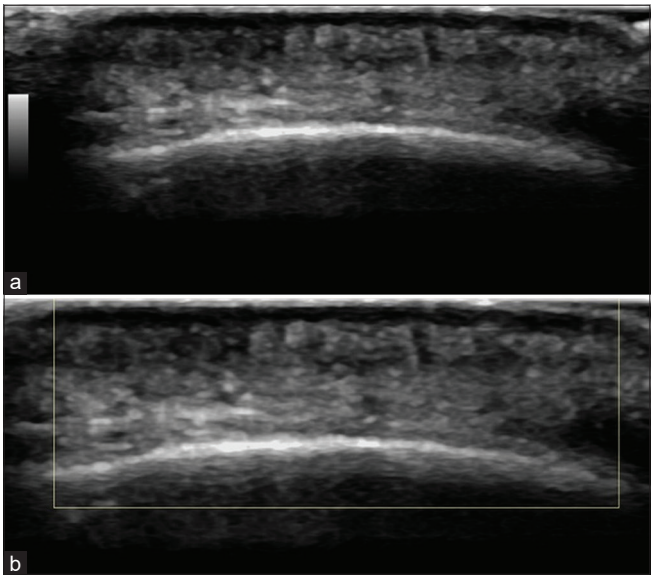

Figure 2: Ultrasound of the scalp region (a) demonstrating an ill defined, hyperechoic lesion occupying the subcutaneous plane of the left scalp area and is not taking any vascularity on color Doppler (b), thus ruling out any underlying vascular malformation.

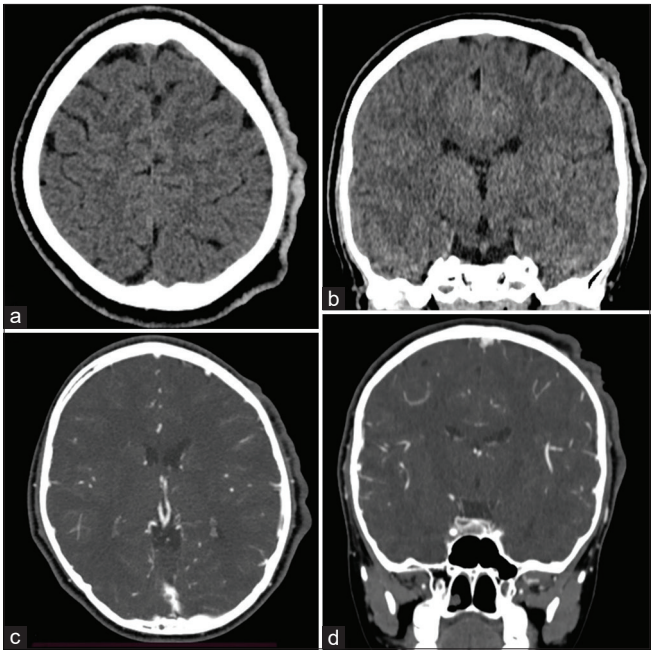

Figure 3: CT brain axial (a) and coronal (b) sections reveal extracranial, irregular, cerebriform pattern of thickening, involving dermal, epidermal, and adipose layers of scalp in left fronto-parieto-temporal regions and post-contrast study (c and d) shows no obvious evidence of feeding arteries/draining veins. Underlying bony cortex appears intact with no features of cortical breach/calvarial thickening.

diagnosis of CVG, secondary to CIN was established and thus confirmed the radiological findings.

DISCUSSION

CVG, also known as cutis verticis plicata/paquidermia verticis gyrata/bulldog scalp syndrome, is a rare skin disorder characterized by ridges and furrows similar in appearance to the whorls of the cerebral cortex.[5] CVG was first described by Jean-Louis-Marc Alibert in 1837and CVG being proposed by Unna in 1907 and remains the accepted name till date. In 1953 Polan and Butter-worth in their review of 195 cases, classified CVG as primary (47.7%) and as secondary (52.3%).

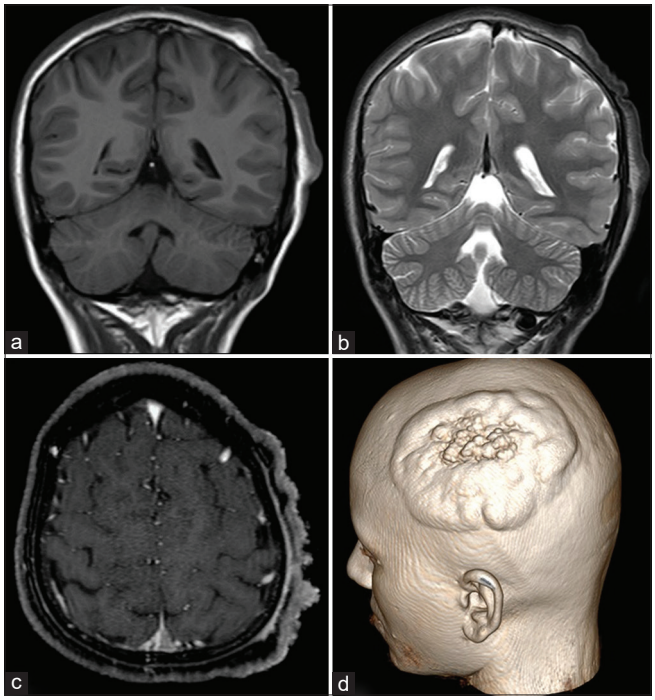

Figure 4: MRI coronal T1 (a) and T2 sequence (b) reveal extracranial, irregular, cerebriform pattern of thickening, involving dermal, epidermal, and adipose layers of scalp and shows subtle post-contrast enhancement (c). The underlying brain parenchyma shows normal enhancement pattern with no intracranial extension. Post-contrast MR 3D surface rendered image of the scalp lesion (d).

CVG is classified into three categories depending on the intrinsic etiology: Primary essential, primary nonessential, and secondary. Primary essential CVG is a solitary finding, with no association with any neurological, endocrine, or ophthalmological disease. Many patients are asymptomatic, and they present for evaluation due to cosmetic reasons. Primary non-essential CVG is typically affiliated with underlying neuropsychiatric disorders, mental retardation, and other brain or ophthalmological conditions. Between 0.2% and 4.5% of patients with primary non-essential CVG have concomitant intellectual disability. Finally, secondary CVG can be related to a variety of other disorders; which include hormonally driven diseases such as acromegaly, thyroid conditions and myxedema; hematologic diseases such as leukemia or amyloidosis; infectious agents such as syphilis or human immunodeficiency virus; inflammatory skin disorders such as eczema, psoriasis or Darier's disease; solitary neoplasms; and genodermatoses such as noonan syndrome, tuberous sclerosis complex, or neurofibromatosis.[6]

The average number of skin folds with CVG in one study was six, though it may vary from two to as many as twenty folds.[7] The direction of the folds is usually anterior to posterior in vertex but may be transverse over the occipital region or sometime can be irregular. In primary CVG, folds are usually symmetrical and are asymmetrical in secondary form.[8]

One of the rarest forms of secondary CVG is the nevoid form, in which it is melanocytic intradermal nevi that cause the cutaneous hypertrophy. This condition is known as CIN and was first linked with CVG in 1937 by Hammond and Ransom.[9] Orkin *et al*. demonstrated that patients with CIN causing CVG have normal intellectual ability with slight female predominance and are free from other local or systemic diseases.[10]

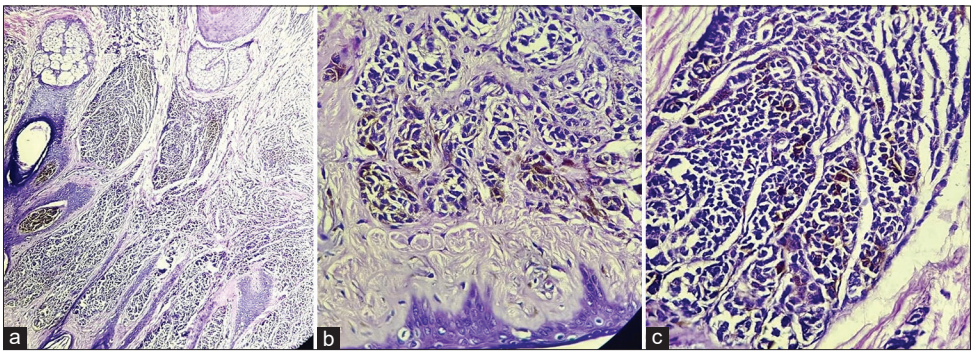

Figure 5: (a-c) Histopathology - Haematoxylin and Eosin (H & E) stain. (x 40). The excised specimen reveals epidermis and dermis with dermis showing in full thickness nevus cells arranged in nests, cords and lying singly. The cells are round to oval with abundant eosinophilic cytoplasm and round to oval nucleus with few of them containing varying amount of melanin along with entrapped adnexal structures - hair follicles, sebaceous glands, and scant inflammatory infiltrates in the dermis.

Histopathologically CIN reveals: The presence of nevocytes within the dermal layer with minimal junctional component, enveloping the adjacent structures with loss of the typical arrangement of nests in stretches. A prevalence of type C nevus cells with large areas of neuroid differentiation; irregular arrangement and distribution of intracellular melanin pigment and perivascular nevocyte aggregation may be appreciated.[11]

Surgical treatment is usually for cosmetic reasons as quality of life may be compromised due to unesthetic aspect. Surgical modalities range from simple excision to tissue expansion and skin grafts for those cases with more extensive scalp involvement.[12]

CIN may evolve into malignant melanoma (MM), which can appear even early in life. Orkin *et al*. reviewed 50 cases of CIN reported up to 1974 and found that two cases of MM were derived from CIN.[10] Currently whether surgical excision is unequivocally required or close follow-up is sufficient is debated upon, given the low rate of malignant transformation and the extensive surgical margins involved with CVG.[9] As MM has been reported in patients with congenital nevus, regular follow-up is necessary in all cases of CIN.

CONCLUSION

CIN presenting as CVG in an adolescent healthy girl is a rarity. Histopathology helps to confirm the diagnosis. Radiological imaging plays a crucial role in characterization of the lesion and helps to rule out any underlying vascular malformation, calvarial involvement, or intracranial extension.

Teaching points

- Cutis verticis gyrate (CVG), is a rare cutaneous disorder, characterized by cerebriform appearance of the scalp skin with convoluted folds and deep furrows (better seen on cross sectional imaging).
- It is usually located on the scalp, but other locations such as neck, lower limbs, buttocks, scrotum, or back have also been reported.
- Classified as primary essential, primary nonessential, and secondary (pseudo CVG) forms.
- The skin folds in CVG are potential source of infection due to accumulation of secretions and may result in fetid or butyric acid odor.
- Mainstay of treatment is educating patient regarding local hygiene care and to avoid secretion accumulation. Surgical treatment is usually for cosmetic reasons as quality of life may be compromised due to unesthetic aspect

MCQs:

1. An adolescent girl with good scholastic performance presented with irregular cerebriform pattern of thickening over her scalp region since birth, which has progressively increased in size to the present condition. She denies any history of trauma, surgery, or any associated conditions with respect to the lesion, what might be the probable diagnosis?
   a. Cutis laxa
   b. Chronic inflammatory condition
   c. Cutis vertisis gyrata
   d. Pachydermoperiostosis.

Answer Key: c

2. Cutis verticis gyrata is also known by these names, except?
   a. Cutis verticis plicata
   b. Paquidermia verticis gyrata
   c. Cutis laxa
   d. Bulldog scalp syndrome.

Answer Key: c

3. True about cutis verticis gyrata of scalp:
   a. Limited to subcutaneous and dermal layers of scalp
   b. Has intra cranial extension
   c. Has distant metastasis
   d. Has laxity of involved scalp area.

Answer Key: a

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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