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A rare case of Giant Cerebriform Intradermal Nevus in a 23 year old male presenting as secondary Cutis Verticis Gyrata

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Abstract

Cutis verticis gyrata is a rare skin condition characterized by ridges and furrows resembling the surface of brain. It manifests as a scalp deformity resembling the surface of the brain, with cerebriform morphologic characteristics. These lesions may progress to malignancy or become infected. Herein we report a rare case of 23 year old male with cerebriform and soft folds on left fronto-parietal areas. On histopathology it showed nests of nevus cells in dermis. The diagnosis of cerebriform intradermal nevus was confirmed. We report this case for its rarity.

Keywords*:* Cutis verticis gyrata, cerebriform intradermal nevus, scalp skin disorder.

Introduction

Cutis verticis gyrata (CVG) is a rare scalp disorder characterized by redundant skin folds. The occurance is said to be 1 in 100,000 in males and 0.026 in 100,000 in females. [1]

CVG is divided into primary and secondary. Primary CVG may be classified into essential CVG, if no other abnormalities are identified, and non-essential CVG, which can be associated with mental deficiency, cerebral palsy, epilepsy, seizures, or ophthalmological disorders. [2] The secondary form of CVG is caused by an underlying condition, such as amyloidosis, syphilis, acromegaly myxedema, pachydermoperiostosis, neurofibroma, giant congenital melanocytic nevus (GCMN) or cerebriform intradermal nevus (CIN). [2]

Cerebriform intradermal nevus was first described by Hammond and Ransom in 1937 and is one of the causes of secondary cutis verticis gyrata.[3] Clinically, cerebriform intradermal nevus is a nonpigmented plaque with sparse hairs in contrast to giant

melanocytic nevus, which is darkly pigmented and hairy.[4] It may also occur in association with various syndromes (Noonan syndrome, Beare–Stevenson syndrome, Ehlers–Danlos Syndrome, "Michelin tire baby" syndrome, Turner syndrome, and fragile X syndrome).[2,5]

CIN usually starts as a flat macule that transforms into convolutions, occupying one half to three quarters of the parietal and occipital scalp. [6, 7] It has a growth peak at puberty. The mechanism for the cerebriform pattern on the surface of the intradermal cerebriform nevus is still unknown.

Case Report

A 23-year-old man presented to our dermatology outpatient for a large convoluted skin lesions over the scalp that started as a small brown colored spot at birth. It gradually grew over the years but remained asymptomatic. Patient's medical and familial history was unremarkable. Cutaneous examination revealed a soft skin-colored folded mass extending from the left fronto-parietal region to the cranial vertex with normal overlying skin, and sparse hair growth (Figure 1a and 1b). Patient's general examination was unremarkable.

Initial clinically diagnosis was kept as secondary cutis verticis gyrata. Biopsies from different sites of the lesion were done, all came with similar histopathological features deep seated hair follicles and nests of nevus cells in the dermis (Figure 2a). It was possible to observe groups of nevus cells forming sparse nests that were diffusely distributed in the dermis (Figure 2b). These cells were related to appendages, as in congenital nevus, with some clear spaces (Figure 2c). These clear spaces were composed of mucopolysaccharides and that there were enlarged capillaries, possibly explaining the progressive growth of the affected area. Routine blood tests along with ophthalmological and neurological referral were all within normal limits. We reported this case for its rarity.

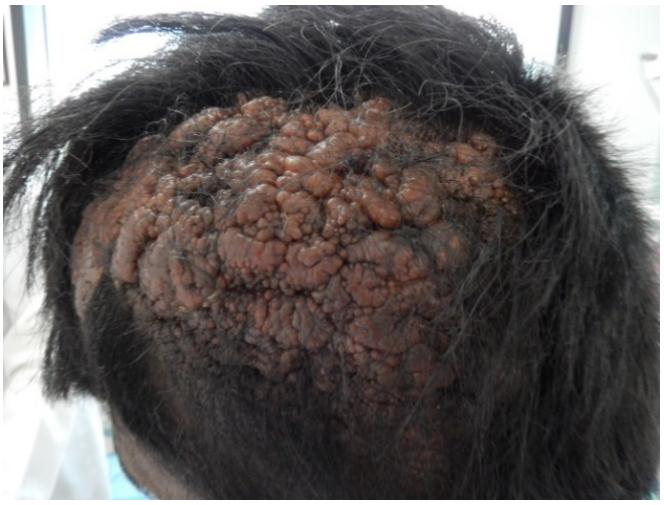

Figure 1a: soft skin-colored folded mass extending from the left fronto-parietal region to the cranial vertex with normal overlying skin, and sparse hair growth

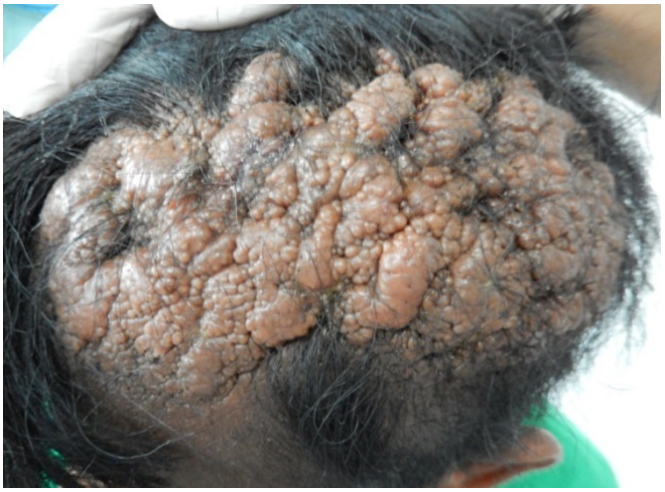

Figure 1b: soft skin-colored folded mass extending from the left fronto-parietal region to the cranial vertex with normal overlying skin, and sparse hair growth

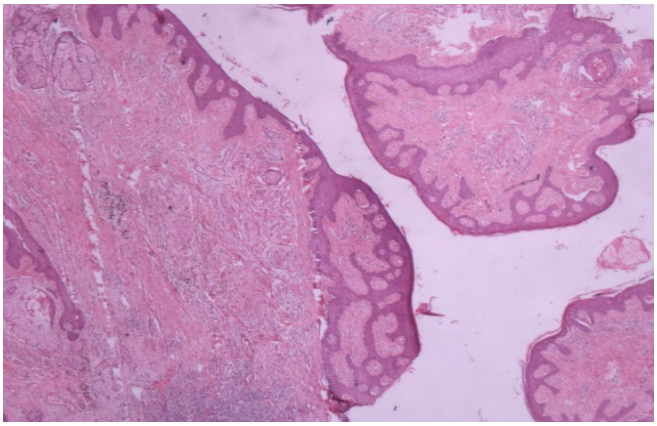

Figure 2a: The epidermis is unremarkable. Dermis show nests of uniform cuboidal to polygonal cells. (H and E, ×4)

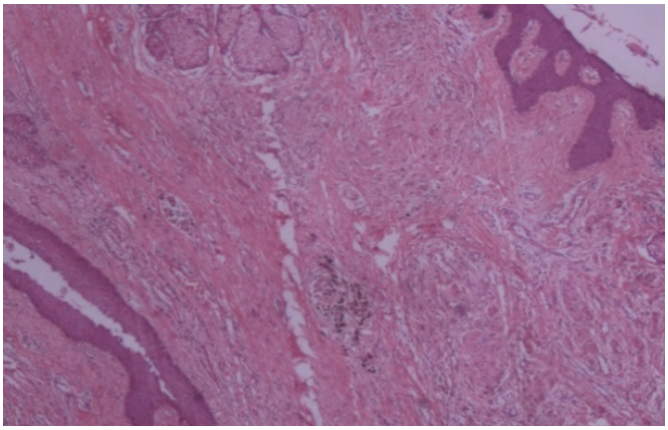

Figure 2b: Nests of nevus cells in the papillary and deep reticular dermis without any junctional component and cellular atypia. Sparse hair follicles in the dermis (H and E, ×10)

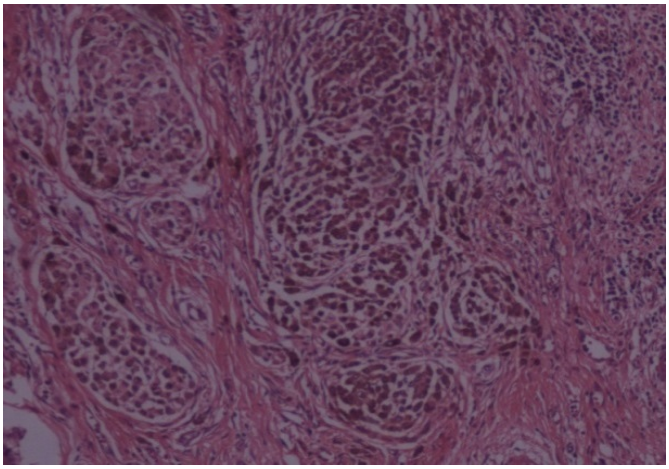

Figure 2c: Deep dermis shows islands of nevus cells with cytoplasmic granular brown pigmentation. No atypical features No features of a deposition disorder identified. (H & E x100)

Discussion

Cerebriform intradermal nevus is a rare cause of secondary CVG. [8, 9] It usually presents at birth or early life as convulations over scalp. Cerebriform intradermal nevus appears as an asymmetric, skin colored, or slightly pigmented tumor that is usually localized in the parietal or occipital areas of the scalp. [9] Over the years, the nevus slowly enlarges and becomes more prominent. Pregnancy, surgical exploration, and hormonal activity have been related to the growth of these lesions. [10]

Histological examination reveals intradermal nevus cells present in the dermis. The nests of nevus cells can be well delineated or irregular. Neuroid transformation can be seen in the deeper parts of the lesion with increase in collagen fibers. Hair follicles can be normal or atrophic. [4,8,10]

Cerebriform intradermal nevus and giant congenital melanocytic nevus are thought to be related conditions by some authors. They share similar histopathologic features, but they have several differences. Giant congenital melanocytic nevus has intense pigmentation, an increased number of hair follicles and frequent nevus cell nests at the dermoepidermal junction, whereas cerebriform intradermal nevus is usually a slightly or non-pigmented lesion, with absent or sparse hair follicles, and uncommon junctional activity [8].

Rarely progression to melanoma has been reported to occur in giant cerebriform intradermal nevus [4]. Because of the risk of malignant transformation and for cosmetic reasons surgical excision and plastic reconstruction are frequently performed. [11] When they are not possible, close follow-up is mandatory.

References

1. Snyder MC, Johnson PJ, Hollins RR. Congenital primary cutis verticis gyrata. Plast Reconstr Surg. 2002;110(3):818-821.
2. Larsen F, Birchall N. Cutis verticis gyrata: three cases with different aetiologies that demonstrate the classification system. Australas J Dermatol. 2007 May;48(2):91-4.
3. Von Geest AJ, Berretty PJ, Klinkhamer PJ, Neumann HA. Cerebriform intradermal naevus (a rare form of secondary cutis verticis gyrata). J Eur Acad Dermatol Venereol. 2002;16:529-31.
4. Tagore KR, Ramineni AK. A case of cutis verticis gyrata secondary to giant cerebriform intradermal nevus. Indian J Pathol Microbiol 2011;54:624-5.
5. Schöttler L, Denisjuk N, Körber A, Grabbe S, Dissemond J. Asymptomatic cerebriform folds of the scalp. Diagnosis: primary Cutis verticis gyrata (CVG). Hautarzt. 2007;58:1058–1061.
6. Polan S, Butterworth T. Cutis verticis gyrata: review with report of seven new cases. Am J MentDefic. 1953; 57:613-631.
7. Corbalan-Velez R, Perez-Ferriols A, Aliaga-Bouiche A. Cutis verticis gyrata secondary to hypothyroid myxedema. Int J Dermatol. 1999; 38:781-783.
8. Yazici AC, Ikizoglu G, Baz K, Polat A, Ustunsoy D. Cerebriform intradermal nevus. Pediatr Dermatol. 2007;24:141–3.
9. Tabata H, Yamakage A, Yamazaki S. Cerebriform intradermal nevus. Int J Dermatol. 1995;34:634.
10. Alcántara González J, Truchuelo Díez MT, Carrillo Gijon R, Martin Diaz RM, Jaen Olasolo P. Letter: Cerebriform intradermal nevus presenting as secondary cutis verticis gyrate. Dermatol Online Journal. 2010;16:14.
11. Goil, P., Jain, A., Patil, A.N. *et al.* Surgical management of giant congenital melanocytic nevus of face in the adult—outcomes and experience. *Eur J Plast Surg* 41, 529–534 (2018).