Cutis Verticis Gyrata Secondary to a Cerebriform Intradermal Nevus

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We report the case of a 30-year-old black man with a large mass consisting of longitudinal parallel ridges and furrows on the left parietal region. A small, single, hyperpigmented macule was present at birth and gradually grew and extended over the years. Cutis verticis gyrata was suspected, and an investigation was performed to discharge the possibility of pachydermoperiostosis. However, results of biopsies obtained from 3 different lesional areas showed the same histopathologic features—deep-seated hair follicles and clusters of nevus cells concentrated in the dermis. Cerebriform intradermal nevus is a rare cause of cutis verticis gyrata. Early diagnosis is extremely important to prevent the development of malignant melanoma.

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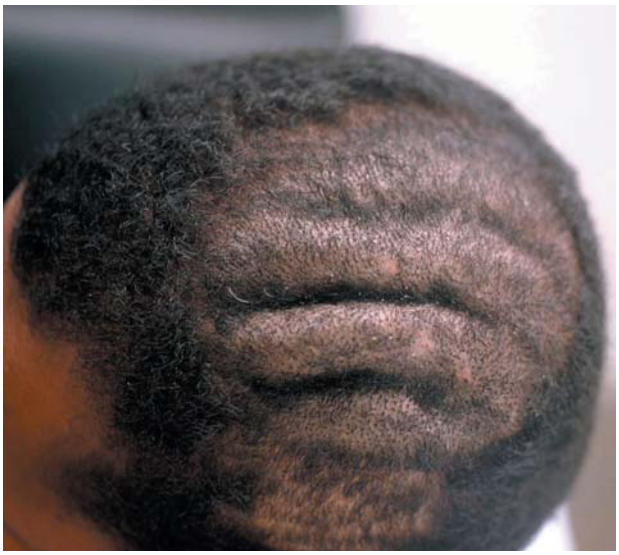

Figure 1. Aspect of the lesion showing the convolutions of the left side of the scalp.

Case Report

A 30-year-old black man consulted our department because of a large tumor on the scalp that presented as a small black spot at birth. Over the years, it gradually grew and extended in a wavelike pattern. The lesion was asymptomatic, and the patient's medical and familial history were unremarkable.

Results of a dermatologic examination revealed a convoluted skin-colored mass extending from the left parietal region to the cranial vertex with soft consistency, normal aspect of the overlying skin, and normal hair growth (Figure 1). There were no symptoms, and results of the patient's general examination were normal.

The clinical diagnosis was cutis verticis gyrate. Punch biopsies from 3 different sites of the lesion were performed, all with the same histopathologic features—deep-seated hair follicles, as expected in a scalp biopsy, and nests of nevus cells in the dermis. It was possible to observe groups of nevus cells forming sparse nests that were diffusely distributed in the dermis. These cells were related to appendages, as in congenital nevus, with some clear spaces (Figure 2). Colloidal iron stain showed that these clear spaces were composed of mucopolysaccharides and that there were enlarged capillaries, possibly explaining the progressive growth of the affected area (Figure 3).

The patient was thoroughly examined by the ophthalmology and neurology sectors without any signs of alterations. Results of routine blood tests also were within reference range.

Comment

Primary cutis verticis gyrata is a rare condition mimicking the surface of the brain, more often affecting young men after puberty (men to women ratio, 5–6:1).[1,2] The usual presentation is composed of 2 to 20 symmetric convolutions on the scalp,

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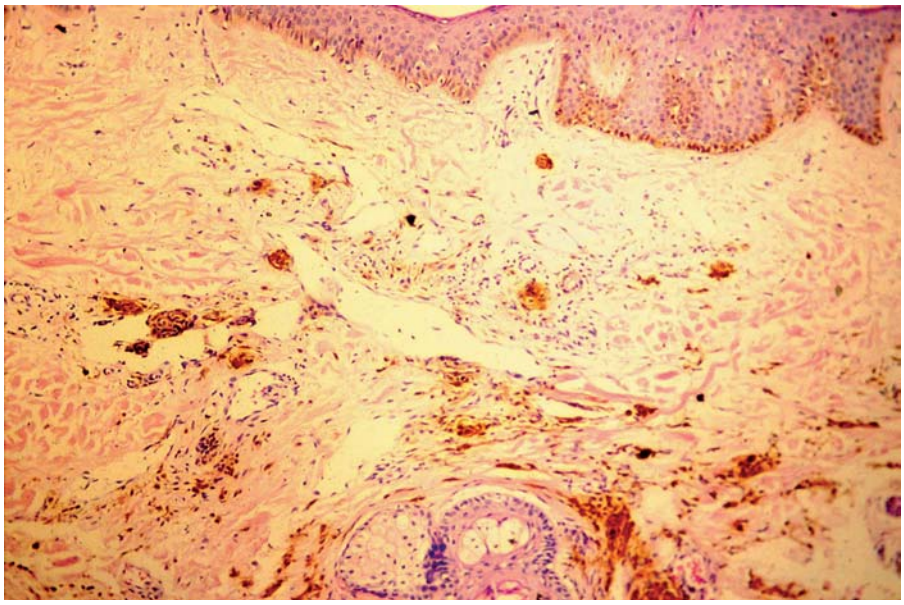

Figure 2. Groups of nevus cells forming nests, sparsely and diffusely distributed in the dermis. These cells were related to appendages, as in congenital nevus (H&E, original magnification ×40).

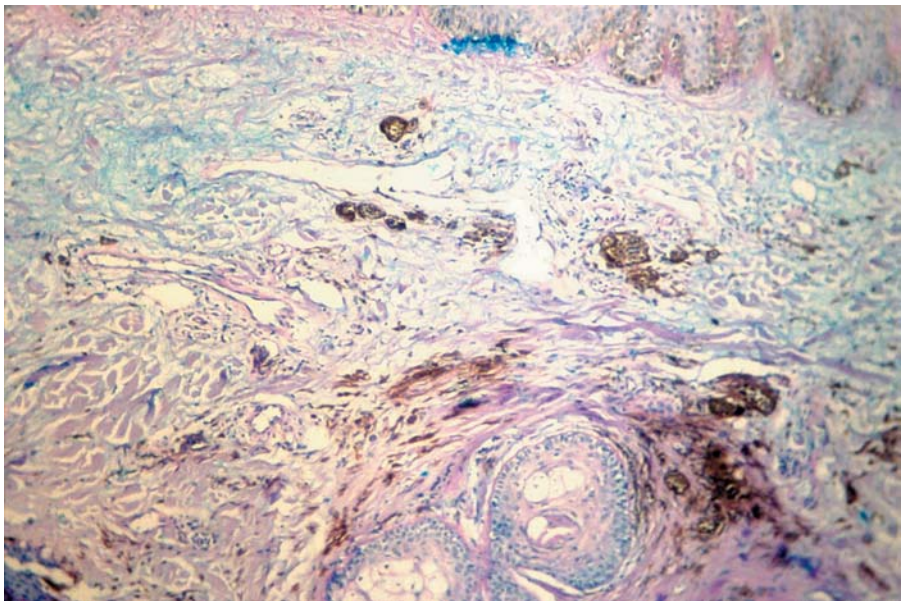

Figure 3. Clear spaces composed of mucopolysaccharides and enlarged capillaries, possibly explaining the progressive growth of the affected area (colloidal iron, original magnification ×40).

directed from the anterior to the posterior region, but it also can manifest on other areas of the body. It is asymptomatic and slow growing and is divided into 2 forms: the primary essential form, associated with neurologic disorders (epilepsy, microcephaly), ophthalmologic disorders (cataract, strabismus, blindness), and/or psychiatric alterations (mental retardation, schizophrenia); and the primary nonessential form, which affects otherwise normal individuals and is much more uncommon.[1,3-5] Secondary, or pseudo, cutis verticis gyrata is related to many disorders including those listed in the Table.[1-3,5-10]

Tumors make up almost 25% of the causes of cutis verticis gyrata. An underlying dermal nevus is the tumor most often found.[11] Intradermal cerebriform nevus is a rare entity, predominantly affecting females at birth or during the first years of life. The condition usually begins with a macular lesion that develops into parietal and occipital symmetric convolutions, occupying one half to three quarters of the 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 unknown.[12]

Cutis verticis gyrata is not associated with systemic diseases, and it has not been shown to impair

Causes of Secondary Cutis Verticis Gyrata[1-3,5-10]

- Acanthosis nigricans
- Acromegaly
- Acute and chronic inflammatory dermatoses
- Amyloidosis
- Chronic traction—trauma
- Ehlers-Danlos syndrome
- Fallopian tube cancer
- Leukemia
- Mucinosis
- Myxedema and hypothyroidism
- Pachydermoperiostosis
- Syphilis
- Tuberous sclerosis
- Tumors (intradermal nevus, lipomatous nevus, sebaceous nevus, neurofibroma, dermatofibroma, hamartoma, cylindroma, histiocytofibroma, lymphangioma)

intelligence.[3,6] However, it may present with alopecia, bleeding, itching, fetid scent, and infection. The diagnosis is based on clinical suspicion and the histopathology of the lesion, which shows numerous nests and isolated nevus cells through the dermis with variable amounts of melanin, all within a collagenous reticular stroma. Hair follicles may appear atrophied and intense vascular proliferation has been reported.[2] This nevus is usually stable, though it can degenerate into malignant melanoma; sometimes it is necessary to perform a lymph node biopsy to discharge this possibility.[1,2,5,6] Cerebriform intradermal nevus is a type of large congenital nevus and should be evaluated with this in mind.[11] In the present case, we demonstrated an extensive deposition of interstitial mucopolysaccharides, which may help to explain the cerebriform clinical aspect.

Treatment consists of a wide excision of the whole tumor and the use of complex flaps. Tissue expansion for coverage of the large area may give better final results, minimizing the esthetical deformity.[1,2,4,5]

This is an extremely rare case and the possibility of evolution into melanoma in 4% to 10% of patients shows the absolute need and extreme importance of early diagnosis of this type of nevus.[1]

REFERENCES

1. Jeanfils S, Tennstedt D, Lachapelle JM. Cerebriform intradermal nevus. a clinical pattern resembling cutis verticis gyrata. *Dermatology*. 1993;186:294-297.
2. Cribier B, Lipsker D, Mutter D, et al. Pachydermie verticellée occipitale: traitement chirurgical de réduction. *Ann Dermatol Venereol*. 1993;120:542-545.
3. Berbis P, Dor AM, Niel-Bourrelly RM, et al. Naevus cérébriforme intra-dermique. *Ann Dermatol Venereol*. 1987;114:369-373.
4. Hamm JC, Argenta LC. Giant cerebriform intradermal nevus. *Ann Plast Surg*. 1987;19:84-88.
5. Hashimoto I, Urano Y, Nakanishi H, et al. Cerebriform intradermal nevus: a case of scalp expansion on the galea. *J Dermatol*. 1999;26:258-263.
6. Orkin M, Frichot BC III, Zelickson AS. Cerebriform intradermal nevus. a cause of cutis verticis gyrata. *Arch Dermatol*. 1974;110:575-582.
7. van Geest AJ, Berretty PJ, Klinkhamer PJ, et al. Cerebriform intradermal naevus (a rare form of secondary cutis verticis gyrata). *J Eur Acad Dermatol Venereol*. 2002;16:529-531.
8. Polan S, Butterworth T. Cutis verticis gyrata: review with report of seven new cases. *Am J Ment Defic*. 1953;57:613-631.
9. Pachinger W, Honig D. Zum krankheitsbild der cutis verticis gyrata. *Z Hautkr*. 1981;56:275-280.
10. Corbalan-Velez R, Perez-Ferriols A, Aliaga-Bouiche A. Cutis verticis gyrata secondary to hypothyroid myxedema. *Int J Dermatol*. 1999;38:781-783.
11. Lasser AE. Cerebriform intradermal nevus. *Pediatr Dermatol*. 1983;1:42-44.
12. Tabata H, Yamakage A, Yamazaki S. Cerebriform intradermal nevus. *Int J Dermatol*. 1995;34:634.