

CASE REPORT

CUTIS VERTICIS GYRATA SECONDARY TO A CEREBRIFORM INTRADERMAL NEVUS

Manisha Nijhawan¹, Shifa Yadav², Aakanksha Singh³, Dinesh Mathur⁴, Puneet Agarwal⁵, Vibhor Goyal⁶, Shilpa Soni⁷,

HOW TO CITE THIS ARTICLE:

Manisha Nijhawan, Shifa Yadav, Aakanksha Singh, Dinesh Mathur, Puneet Agarwal, Vibhor Goyal, Shilpa Soni, "Cutis verticis gyrata secondary to a cerebriform intradermal nevus". *Journal of Evolution of Medical and Dental Sciences* 2013; Vol. 2, Issue 52, December 30; Page: 10332-10335.

ABSTRACT: Cerebriform intradermal nevus is a rare form of cutis verticis gyrata. Clinically it manifests as a scalp deformity resembling the surface of the brain, with cerebriform morphologic characteristics these lesions may become malignant or infected. Here we report a 20 year old boy, with cerebriform and soft fold on right temporo-parietal area present since birth. The clinical and histopathological (solitary or clusters of nevus cells) presentations of cerebriform intradermal nevus are described and the therapeutic possibilities discussed.

INTRODUCTION: Cutis verticis gyrata (CVG) is a skin deformity characterized by excessive growth of the skin of the scalp, resulting in furrows and folds which are similar to gyri of the brain cortex. CVG can be classified into two forms: 'primary' (essential and non-essential) and 'secondary'. The primary non-essential type is associated with the neurological and ophthalmological changes, while the primary essential form does not present any other disorder except for the cutaneous alterations. Secondary forms of CVG are much more frequent and accompany other pathologies. The treatment consists of local hygiene and surgical resection of skin excess. 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

CASE REPORT: A 20-year-old man consulted our department for a large tumor on the scalp that started as a small black spot at birth. Over the years, it gradually grew however remained asymptomatic. Patient's medical and familial history were unremarkable. Results of dermatologic examination revealed a soft skin-colored mass extending from the right temporoparietal region to the cranial vertex with normal overlying skin, and normal hair growth (fig 1). Patient's general examination was unremarkable.

Patient was clinically diagnosed as cutis verticis gyrate. Punch biopsies from 3 different sites of the lesion were performed, all with similar histopathological 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). Routine blood test along with ophthalmology and neurology referral did not reveal any abnormality.

CASE REPORT



Fig. 1

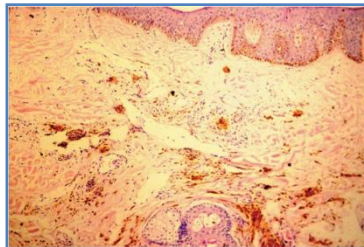


Fig. 2: Groups of nevus cells forming nest

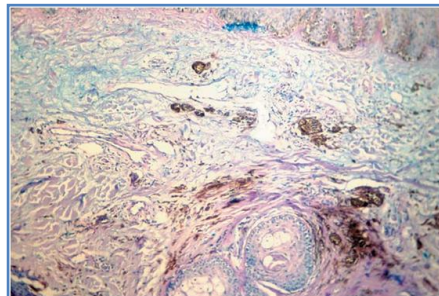


Fig. 3: Clear spaces composed of mucopolysaccharides and enlarged capillaries, possibly explaining the progressive growth of the affected area (colloidal iron, original magnification $\times 40$)

DISCUSSION: Primary cutis verticis gyrata is a rare condition mimicking the surface of the brain,

CASE REPORT

more often affecting young men after puberty (men to women ratio, 5–6:1).¹ This usually presents as two to twenty symmetric convolutions on the scalp, directed from the anterior to the posterior region, though other areas of the body can also be involved. It is an asymptomatic and slow growing and is divided into two 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.²⁻³ Secondary, or pseudo, cutis verticis gyrata is related to many disorders including those listed in (table 1).

Acanthosis Nigricans
Acromegaly
Acute and Chronic Inflammatory dermatoses
Amyloidosis
Chronic traction trauma
Ehler-Danlos Syndrome
Mucinosis
Tumours: Intradermal nevus, Lipomatous Nevus, Sebaceous Nevus, Neurofibroma, Dermatofibroma, Hamartoma, Cylindroma, Lymphangioma
Causes of Secondary Cutis Verticis Gyrata

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. This nevus is usually stable, though rarely it can degenerate into malignant melanoma.³⁻⁴ Cerebriform intradermal nevus is a type of large congenital nevus and should be evaluated with this in mind¹¹

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.²⁻³

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 nevus¹.

REFERENCES:

1. Jeanfils S, Tennstedt D, Lachapelle JM. Cerebriform intra- dermal nevus. a clinical pattern resembling cutis verticis gyrata. *Dermatology*. 1993; 186:294-297.
2. Hamm JC, Argenta LC. Giant cerebriform intradermal nevus. *Ann Plast Surg*. 1987; 19:84-88.
3. 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.
4. Orkin M, Frichot BC III, Zelickson AS. Cerebriform intradermal nevus. a cause of cutis verticis gyrata. *Arch Dermatol*. 1974; 110:575-582.

CASE REPORT

5. vanGeest 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.
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. Lasser AE. Cerebriform intradermal nevus. *Pediatr Dermatol.* 1983; 1:42-44.
9. Tabata H, Yamakage A, Yamazaki S. Cerebriform intra- dermal nevus. *Int J Dermatol.* 1995; 34:634.

AUTHORS:

1. Manisha Nijhawan
2. Shifa Yadav
3. Aakanksha Singh
4. Dinesh Mathur
5. Puneet Agarwal
6. Vibhor Goyal
7. Shilpa Soni

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Skin and VD, Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.
2. Resident, Department of Skin and VD, Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.
3. Resident, Department of Skin and VD, Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.
4. Head Of the Department, Department of Skin and VD, Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.

5. Resident, Department of Skin and VD, Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.
6. Resident, Department of Skin and VD, Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.
7. Resident, Department of Skin and VD, Mahatma Gandhi Medical College & Hospital, Sitapura, Jaipur.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Shifa Yadav,
112, Panchsheel Enclave,
JLN Road, Jaipur.
Email – m.nijhawan12@gmail.com

Date of Submission: 31/10/2013.

Date of Peer Review: 02/11/2013.

Date of Acceptance: 15/11/2013.

Date of Publishing: 27/12/2013.