

Cerebriform patterns in dermatology

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ABSTRACT

Dermatology has vast spectrum of mucocutaneous manifestations in respect of morphology of the lesions, their arrangement, distribution and other feature. Cerebriform pattern is one of many interesting presentation which has been described in some skin conditions in English literature. Here in this article, those conditions have been compiled and described in brief.

Key words: Cerebriform; sulci and gyri, brain like; atypical lymphocytes

INTRODUCTION

Dermatology is a rapidly evolving branch of medicine which has wide spectrum of mucocutaneous manifestations. Morphology of lesions, their arrangement, distribution and associated features help to narrow down the differential diagnosis. There are many academic tools such as mnemonics, acronyms, eponyms, triads, phenomena, etc which make it easier to remember various diagnostic features. Cerebriform appearance is one such peculiar feature which has been described in literature in some dermatoses. Here, we have tried to compile such conditions and segregated them based on the method of examination which revealed them.

Clinical, histopathologic, culture (macroscopic), dermoscopic and reflectance confocal microscopy (RCM) are 5 such examination tools which have demonstrated cerebriform patterns in previous English literature. We have searched standard text books and databases of PUBMED, SCOPUS, INDEX MEDICUS, MEDLINE and Google Scholar. They all have been arranged alphabetically in this article and described concisely focussing on their cerebriform appearance. Of note, large plaque parapsoriasis, lichen planus-like keratosis and lichen sclerosus et atrophicus have seldom demonstrated cerebriform nuclei therefore only mentioned in the Table 1 with no description in the text.

1. Actinic reticuloid – It is commonly known as chronic actinic dermatitis and presents as scaly, indurated, hyperpigmented papules and lichenified plaques, mostly on sun-exposed areas. Histopathology reveals dense pandermal lymphohistiocytic infiltrates along with multinucleate stellate myofibroblasts and giant cells. Large, atypical, hyperchromatic, cerebriform lymphoid cells are also present which can mislead the diagnosis as mycosis fungoides [1].
2. Adult T cell leukemia-lymphoma – It is caused by human retrovirus HTLV-1 which also simulates the clinic-histopathologic pictures of mycosis fungoides but has peripheral blood and significant systemic involvement. On cutaneous histopathology, cellular atypia may be mild but Pautrier-like microabscesses and cerebriform or multilobulated nuclei are prominent features [1].
3. Cerebriform intradermal nevus (CIN) – It is a rare congenital nevus which typically presents as asymptomatic, pigmented cerebriform tumor on the parietal or occipital surface of the scalp [2]. With age, its size and convolutions increase along with progressive loss of hairs (tufts of hair are present on sulci only) and often called as cutis verticis gyrata (Fig. 1).
4. Connective tissue nevus – It may have various morphologic presentations involving dermal collagen (collagenoma) or elastic fibres (elastoma). Both, syndromic and non-syndromic forms

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Table 1: Various types of cerebriform patterns

Clinical	Histopathology	Culture	RCM	Dermoscopy
1. Cerebriform intradermal Nevus	1. Actinic reticuloid	1. Trichophyton tonsurans	1. Malignant melanoma	1. Seborrheic keratosis
2. Connective tissue nevus	2. Adult T cell leukemia-lymphoma	2. Trichophyton schoenleinii		2. Epidermolytic acanthoma
3. Cutis Verticis Gyrata	3. Lymphomatoid drug reaction	3. Paracoccidioidomycosis		
4. Diffuse Smooth muscle hamartoma	4. Lymphomatoid papulosis			
5. Dissecting cellulitis of scalp	5. MF/Sezary syndrome			
6. Nevus lipomatosus Cutaneous Superficialis	6. Pagetoid reticulosis			
7. Nevus sebaceous	7. Granulomatous slack skin			
8. Pemphigus vegetans	8. Large plaque parapsoriasis			
9. Pitted Keratolysis	9. Lichen planus-like keratosis			
10. Verruciform xanthoma	10. Lichen sclerosis et atrophicus			

RCM: Reflectance confocal microscopy

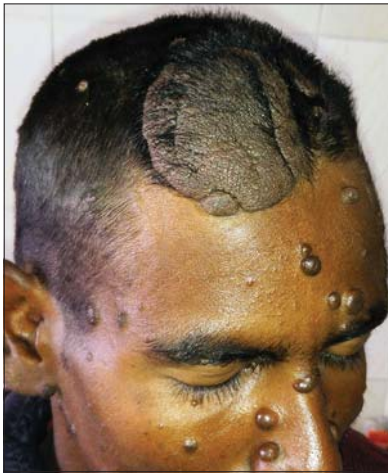


Figure 1: Cerebriform intradermal nevus on scalp and discrete intradermal nevi on face.



Figure 2: Tuberous sclerosis with facial angiobfibroma (under treatment with electrocautery) and forehead fibrous plaque.

are known. Collectively, it usually presents as asymptomatic, skin-colored, non-scaly papules and/or plaques mostly on back, buttocks and extremities in non-syndromic sporadic forms. Cerebriform morphology of collagenoma on sole is a well known diagnostic finding in Proteus syndrome, called as plantar cerebriform collagenoma [3]. Such morphologies can also be seen in shagreen patch and fibrous cephalic plaque in tuberous sclerosis (Figs. 2 and 3).

- Cutis Verticis Gyrata - It is characterized by excessive soft, spongy foldings of the scalp skin showing ridges and furrows similar to the brain. It can be primary (idiopathic) with or without neurologic or ophthalmologic anomalies, or secondary, caused by an underlying condition such as cerebriform intradermal nevus, neurofibroma, pachydermoperiostosis, myxedema, amyloidosis, acromegaly, Turner syndrome, Noonan syndrome, giant congenital melanocytic nevus, cerebriform epithelial connective tissue hamartoma, cylindroma, etc [4].
- Diffuse Congenital Smooth muscle hamartoma – Congenital smooth muscle hamartoma typically presents as localized, variably hyperpigmented, irregular plaque with prominent vellus hairs. In



Figure 3: Fibrous cephalic plaque showing sulci and gyri like pattern.

its diffuse form, there is extensive involvement of extremities leading to Michelin tyre baby appearance and cerebriform palmar surface [5].

- Dissecting cellulitis of the scalp (perifolliculitis capitis abscedens et suffodiens or Hoffman disease) – It is an uncommon cause of scarring alopecia having chronic, relapsing course. There are multiple pustules, fluctuating nodules and interconnecting

- abscesses mostly developing on the vertex which has boggy and sometimes cerebriform pattern [6].
8. Epidermolytic acanthoma – It is a benign, acquired, underdiagnosed entity which typically presents as hyperkeratotic papule/s on trunk and limbs. Dermoscopy often shows pearly white areas (globular, linear and arboriform patterns), cerebriform ridges and peripheral pigmented radial streaks corresponding to the invaginations of the stratum corneum [7].
 9. Lymphomatoid drug reactions – There are some drug reactions (due to anticonvulsants, griseofulvin, cyclosporine, atenolol, etc) which simulate the histopathology of mycosis fungoides. Atypical lymphoid cells with cerebriform nuclei and epidermotropism are its characteristic features [8].
 10. Lymphomatoid papulosis – It is a CD30+ lymphoproliferative disorder characterized chronic, recurrent, self-healing, papulonodular or papulonecrotic lesions. Owing to its clinical similarity with many conditions, histopathologic confirmation is mandatory. It has 3 well recognized histological subtypes: Type A consisting of large atypical CD30+ cells along with small lymphocytes, histiocytes and neutrophils; Type B with epidermotropic infiltrate of small atypical lymphoid cells with cerebriform nuclei resembling mycosis fungoides; and Type C with few admixed inflammatory cells mimicking anaplastic large T-cell lymphomas [9].
 11. Malignant melanoma – On reflectance confocal microscopy (RCM), melanomas with a dermal component show confluent aggregates of low reflecting cells in the papillary dermis separated by a darker rim, giving multilobate cerebriform appearance. It is one of the 4 minor RCM criteria for melanoma with about 93% sensitivity [10].
 12. Mycosis fungoides (MF)/Sezary syndrome and its variants – The leukemic phase of mycosis fungoides is known as Sézary syndrome. The typical tumor cells of this spectrum of cutaneous T cell lymphoma are known as mycosis cells or sezary cells (Lutzner cells for smaller forms with 8–11 μm diameter). These cells have a variable amount of non-granular cytoplasm, convoluted or cerebriform nuclei with intertwined lobes and condensed chromatin (Fig. 4) [1]. Pagetoid reticulosis and granulomatous slack skin are two other variants of MF which differ in clinical presentations but frequently have cerebriform nuclei within atypical lymphocytes.
 13. Nevus Lipomatosus Cutaneous Superficialis – It is a hamartomatous condition of disputed origin,

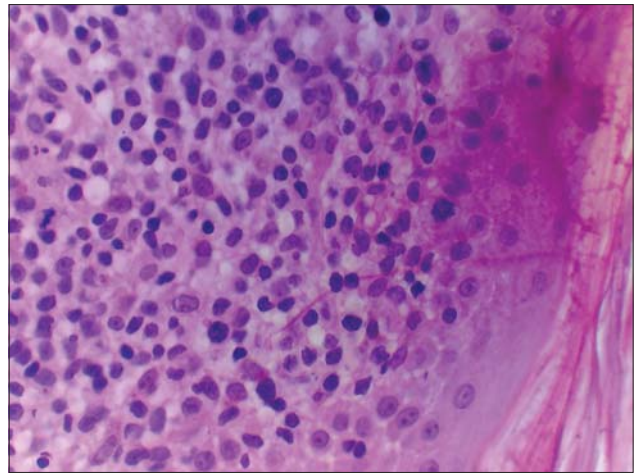


Figure 4: Convoluted, cerebriform nuclei within atypical lymphocytes in MF or sezary syndrome (H & E, x 400).

- characterized by ectopic, mature adipose tissue in the dermis. Mostly, it presents as asymptomatic, soft, skin colored or yellowish plaques with wrinkled or cerebriform surface [11]. Pelvic and lumbar region, and buttocks are the common sites.
14. Nevus sebaceous – It is also called as organoid nevus, mostly presents as a single, yellowish, hairless plaque on the scalp or forehead, at birth or in early childhood. After puberty, its surface becomes verrucous and thickened. Giant cerebriform nevus sebaceous is a recognized atypical presentation which can coexist with other benign appendageal tumors [12].
 15. Paracoccidioidomycosis – It is a deep fungal infection mostly involving lung, lymph nodes, visceral organs and mucocutaneous sites, endemic in South and Central America. It is commonly caused by dimorphic *paracoccidioides brasiliensis*. On culture at 35-37 degree celcius temperature, it characteristically forms creamy, cerebriform or coral-like macroscopic colonies of multiple budding yeasts which looks like mariner’s wheel on microscopy [13].
 16. Pemphigus vegetans - Cerebriform tongue, a typical morphologic pattern consisting of sulci and gyri over dorsum of the tongue is a well-known ‘*Premalatha sign*’ seen in pemphigus vegetans (Fig. 5) [14]. It can also be seen over the flexures of the body and thus can help in the diagnosis of pemphigus vegetans.
 17. Pitted keratolysis – It is an asymptomatic, superficial skin infection usually caused by *Kytococcus sedentarius*, *Dermatophilus congolensis* and *corynebacterium*. Soles are the sites of predilection and hyperhidrosis is often associated



Figure 5: Cerebriform tongue in pemphigus vegetans.



Figure 6: Pitted keratolysis with cerebriform appearance.

with foot odour. It classically presents as bilaterally symmetrical, multiple, shallow, rounded crateriform pits on weight bearing areas of sole. Sometimes, maceration may occur which may look like cerebriform appearance (Fig. 6) [15].

18. Seborrheic keratosis - Fissures and ridges are diagnostic dermoscopic findings of seborrheic keratosis. These are brownish-black structure with papillomatous surface, giving the appearance of sulci and gyri of brain [16]. Individual sulcus are known as fat fingers.
19. Tinea capitis – It is a dermatophytosis of scalp skin, commonly caused by different species of trichophyton and microsporum. Interestingly, trichophyton tonsurans and trichophyton schoenleinii form cerebriform colonies on culture. The colonies are creamy in T. tonsurans and powdery and brownish in case of T. schoenleinii [17].
20. Verruciform xanthoma – It usually presents as asymptomatic, single, skin-colored, papillary growth which mostly involve oral mucosa and occasionally anogenital region. Only one case presenting as cerebriform papules on scrotum has been reported by Andreychik et al in 2015 [18].

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

REFERENCES

1. Reddy K, Bhawan J. Histologic mimickers of mycosis fungoides: a review. *J Cut Pathol.* 2007;34:519-25.
2. Khanna D, Goel K, Khurana N. Isolated plantar cerebriform collagenoma. *Indian J Dermatol Venereol Leprol* 2012;78:666.
3. Topal IO, Goncu OE, Leblebici C, Gungor S, Duman H, Erdemir AV. Cutis verticis gyrata secondary to giant cerebriform intradermal nevus in a pediatric patient. *Indian J Paediatr Dermatol.* 2017;18:50-2.
4. Phiske M. Cerebriform intradermal nevus: A rare entity and its associations. *Indian Dermatol Online J.* 2014;5:115-6.
5. Glover MT, Malone M, Atherton DJ. Michelin tyre baby syndrome resulting from diffuse smooth muscle hamartoma. *Pediatr Dermatol.* 1989;6:329–31.
6. Mundi JP, Marmon S, Fischer M, Kamino H, Patel R, Shapiro J. Dissecting cellulitis of the scalp. *Dermatol Online J.* 2002;18:8.
7. Behera B, Gochhait D, Sridivya P, Chandana S, Thappa DM, Malathi M. Dermoscopy of a solitary verrucous plaque on the back. *J Am Acad Dermatol.* 2017;77:e37-9.
8. Welsh JP, Ko C, Hsu WT. Lymphomatoid drug reaction secondary to methylphenidate hydrochloride. *Cutis.* 2008;81:61-4.
9. Mani V, George R, Vijayakumar K, Nair S. Type D lymphomatoid papulosis simulating aggressive epidermotropic cytotoxic lymphoma. *Indian J Pathol Microbiol.* 2016;59:81-3.
10. Ahlgrimm-Siess V, Laimer M, Rabinovitz HS, Oliviero M, Hofmann-Wellenhof R, Marghoob AA, et al. Confocal Microscopy in Skin Cancer. *Curr Derm Rep* 2018;7:105–18.
11. Patil SB, Narchal S, Paricharak M, More S. Nevus lipomatosus cutaneous superficialis: a rare case report. *Iran J Med Sci.* 2014;39:304-7.
12. Mahajan R, Dogra S, Kanwar AJ, Saikia UN, Agrawal S. Extensive cerebriform nevus sebaceus: an unusual presentation. *Dermatol Online J.* 2012;18:9.
13. Hahn RC, Rodrigues AM, Della Terra PP, Nery AF, Hoffmann-Santos HD, Go'is HM, et al. Clinical and epidemiological features of paracoccidioidomycosis due to *Paracoccidioides lutzii*. *PLoS Negl Trop Dis.* 2019;13: e0007437.
14. Premalatha S, Jayakumar S, Yesudian P, Thambiah AS. Cerebriform Tongue-a clinical sign in pemphigus vegetans. *Br J Dermatol.* 1981;104:587-9.
15. Fernández-Crehuet P, Ruiz-Villaverde R. Pitted keratolysis: an infective cause of foot odour. *CMAJ.* 2015;187:519.
16. Sonagara B, Mehta H, Astik B, Agrawal N. Study of seborrheic keratosis by dermoscopy using polarized and nonpolarized modes. *Indian J Dermatopathol Diagn Dermatol.* 2020;7:64-9.
17. Surendran K, Bhat RM, Bolor R, Nandakishore B, Sukumar D. A clinical and mycological study of dermatophytic infections. *Indian J Dermatol.* 2014;59:262-7.
18. Andreychik C, Bernstein LJ, Elston D. Dermatopathology quiz: Cerebriform papules of the scrotum. *Indian Dermatol Online J.* 2015;6:416-8.

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