

Cerebriform intradermal nevus presenting as cutis verticis gyrata with multiple cellular blue nevus over the body: A rare occurrence

Somenath Sarkar, Soumyajit Roychoudhury, Arpit Shrimal, Kapildeb Das

Department of
Dermatology,
Venereology and Leprosy,
Calcutta School of
Tropical Medicine,
Kolkata, India

ABSTRACT

Cutis verticis gyrata is a rare skin condition characterized by swelling of scalp resembling the surface of the brain. Various conditions, like cerebriform intradermal nevus (CIN), may give rise to this clinical entity. Moreover, its association with cellular blue nevus is extremely rare and has not been reported so far. Here, we report a 28-year-old male with a huge cerebriform swelling covering the occipital lobe along with multiple nodules all over the body. Histology of the scalp swelling showed solitary or clusters of nevus cells in the dermis and from the body lesions showed features of cellular blue nevus. The diagnosis of CIN with cellular blue nevus was confirmed.

Key words: Cellular blue nevus, cutis verticis gyrata, cerebriform intradermal nevus

INTRODUCTION

Cutis verticis gyrata (CVG) is a rare skin condition characterized by ridges and furrows resembling the surface of the brain.^[1] In general, it occurs over the scalp, but other areas such as neck, back, buttocks, scrotum have also been reported as the site of this lesion.^[2] Various conditions may give rise to CVG and based upon the etiology, it has been divided into primary and secondary types.^[1] Primary types may be either essential or non-essential. Essential types present solely with CVG, but non-essential cases are associated with mental deficiency, cerebral palsy, epilepsy, seizures or ophthalmological disorders.^[3] The secondary form of CVG is caused by underlying conditions such as cerebriform intradermal nevus (CIN),^[4] amyloidosis, syphilis, acromegaly,^[5] myxedema, pachydermoperiostosis,^[6] neurofibroma, giant congenital melanocytic nevus (GCMN).^[2] Although rare, cases of primary and secondary CVG have been reported in the literature, but so far no case has reported its occurrence along with cellular blue nevus. To the best of our knowledge, it is the first case report of co-existence of these two very rare clinical entities.

CASE REPORT

A 28-year-old male patient presented to the skin out-patient department with history of a



Figure 1a: Bosselated irregular non-tender skin colored swelling over the occipital region extending toward the nape of the neck

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.126026

Quick Response Code:



Address for
correspondence:

Dr. Soumyajit
Roychoudhury,
Gokarna,
Murshidabad - 742 136,
West Bengal, India.
E-mail: srcforu@gmail.com

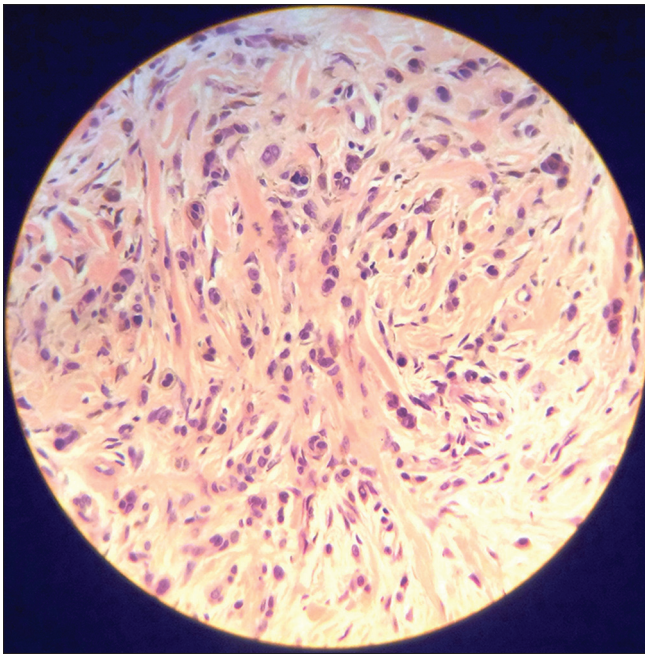


Figure 1b: ×400, H and E showing multiple nests and cords of nevus cells in the upper dermis, which contain a moderate amount of melanin

huge progressively increasing swelling over the back of the scalp [Figure 1a] and multiple black nodules at various parts of the body. According to patient same kind of lesions were present over the trunk at birth, which were surgically removed when he was 4-year-old. The lesion over the scalp started at the age of 6-7 years and the truncal lesions developed gradually since then. On examination, there was a single bosselated irregular non-tender skin colored swelling of 15 cm × 20 cm with deep folds over the occipital region extending toward the nape of the neck. There were certain areas of depigmentation [Figure 1c] along with blue streaks over the swelling. The surface of the skin was hairless, the skin over the swelling was non-pinchable and the swelling was immobile. There was no skin erosion, dermatitis, infection, odor or tenderness.

There were multiple hyperpigmented nodules distributed over different areas of the body such as mandible, flanks, abdomen [Figure 2b], front and back of both thighs, right knee joint, lower leg [Figure 2e], right sole and over volar aspect of first and second finger of the left hand [Figure 2a]. The size of the nodules ranged from 0.5 cm × 1 cm to 1.5 cm × 2.5 cm. The nodules were blackish brown in color surface was smooth and hairless. They were asymptomatic and non-tender on palpation. Nails and mucous membrane examination were within normal limits. He had normal intelligence (the full-scale intelligence quotient was 97) and he had no psychiatric problem. Examination of the neurological, ophthalmological and other systems was normal. Laboratory results, including venereal disease research laboratory test, serology for human immunodeficiency virus, growth hormone and thyroid function,



Figure 1c: Side view of the lesion over scalp

were all negative or normal. A punch biopsy from the scalp lesion and an excision biopsy from the nodule over the body was performed. Section from the specimen showed multiple nests and cords of nevus cells in the upper dermis, which contained a moderate amount of melanin [Figure 1b]. There was no cellular atypia. Section from the body lesion showed deeply pigmented dendritic melanocytes in addition to nests and fascicles of spindle shaped cells with abundant pale cytoplasm containing little melanin [Figures 2c and d]. The spindle cells were arranged in a storiform pattern. Based on the clinical feature and histology it was diagnosed as a case of congenital melanocytic nevus with cellular blue nevi.

DISCUSSION

CVG is a term that refers to a pattern of redundant skin on the scalp that exhibits deep furrows and convolutions. It presents as a skin colored or slightly hyperpigmented swelling with bosselated, smooth surface usually over the occipital and parietal area of the scalp. CIN is a rare cause of secondary CVG.^[2] CIN presents either at birth or during



Figure 2a: Lesions over fingers



Figure 2b: Lesions over flanks, abdomen

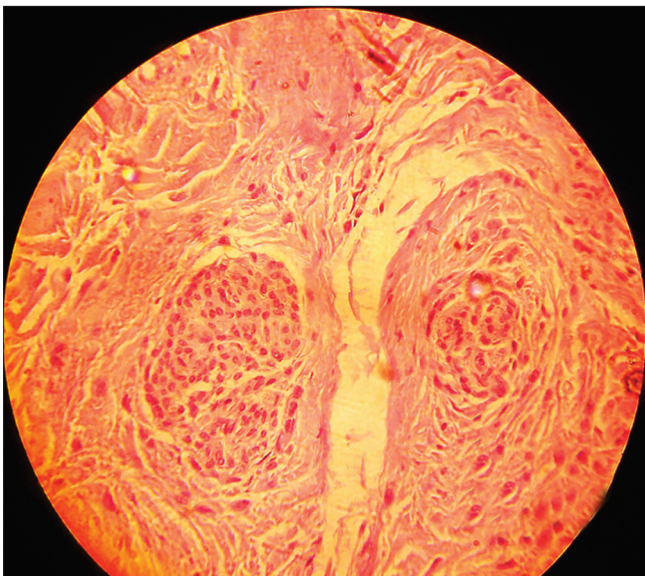


Figure 2c: $\times 400$, H and E stain from the truncal lesion showing nests and fascicles of spindle shaped cells with abundant pale cytoplasm

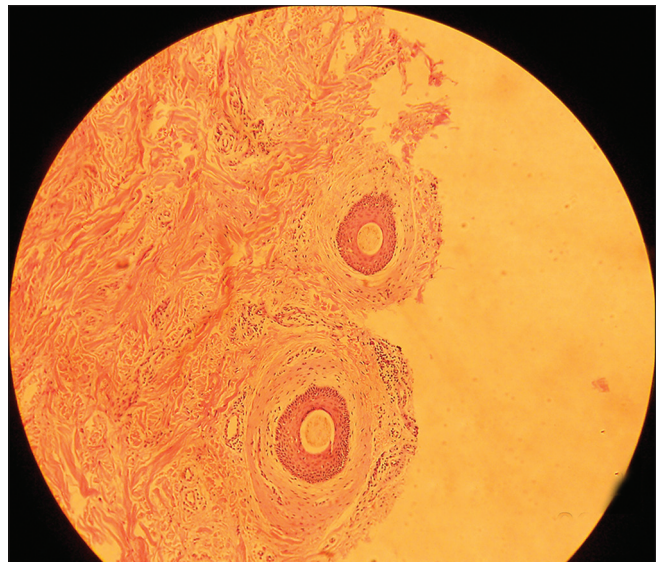


Figure 2d: $\times 100$, view in H and E stain form truncal lesions shows fascicles of spindle shaped cells

early childhood as a gradually enlarging skin-colored or slightly pigmented asymmetric and usually asymptomatic lesion. Lesional alopecia is common. Patients with CIN usually have normal intelligence. A higher incidence of the disease is reported in females.^[7] Pruritus, burning sensation and pain may occur.^[7] Although scalp is the most common site involved, occurrence over the back has recently been reported.^[2]

In our case, CIN presented as skin colored swelling over the occipital region extending toward the nape of the neck leading to CVG pattern. Patient had no sign of any CNS disorder or any ophthalmological disorder. Histology from the scalp lesion confirmed the diagnosis as CIN. Other differential diagnoses that were considered in the present case and ruled out on histological grounds were other causes of secondary CVG,

plexiform neurofibroma, nevus lipomatosus, aggregated cylindromas, connective tissue nevus, amyloid deposition and long-standing dissecting cellulitis of the scalp and multiple metastatic nodules of scalp.

The multiple hyperpigmented nodules distributed over various parts of the body revealed cellular blue nevus on histopathological examination. The differential diagnosis of the nodules was neurofibroma, multicentric reticulohistiocytosis, sarcoidosis, keloids, dermatofibroma. Histologically these entities are different from CIN.



Figure 2e: Lesions over back of legs

The cellular blue nevus was first described as a variant of melanoma. Later, it was classified as a variant of blue nevus. It is composed of the same elements as the common blue nevus, but in addition possessed islands of larger cells arranged in neuroid ("pigmented neurofibroma") or sarcomatoid fashion. Somatic activating mutations in GNA11 and GNAQ (primarily the latter) have been detected in 65-75% of blue nevi.^[8] The genetic basis of coexistence of CIN and cellular blue nevi is not clear.

Rare cases of malignant melanoma have been reported arising in association with cellular blue nevi.^[9]

Because of the rare possibility of malignant transformation, surgical excision and plastic reconstruction of CIN is the preferred treatment option. We referred the case to the

plastic surgeon who advised serial excision of the scalp lesion and flap reconstruction. Blue nevi that are less than 1 cm in diameter, clinically stable, do not have atypical features and are located in a typical anatomic site do not require removal.^[10] Patient was advised six monthly follow-up for the truncal lesions to detect early malignant change, if any.

Our case is unique because, along with the cerebriform nevus over the occiput, there were multiple cellular blue nevi over different parts of the body. Though CIN has been previously reported to present as CVG, blue streaks over the lesion are new in our case.^[11] The rare occurrence of CIN in CVG pattern with multiple cellular blue nevi over the body have made this case a reportable one.

REFERENCES

1. Alcántara González J, Truchuelo Díez MT, Carrillo Gijón R, Martín Díaz RM, Jaén Olasolo P. Letter: Cerebriform intradermal nevus presenting as secondary cutis verticis gyrata. *Dermatol Online J* 2010;16:14.
2. Quaedvlieg PJ, Frank J, Vermeulen AH, Toonstra J, van Neer FJ. Giant cerebriform intradermal nevus on the back of a newborn. *Pediatr Dermatol* 2008;25:43-6.
3. Jeanfils S, Tennstedt D, Lachapelle JM. Cerebriform intradermal nevus. A clinical pattern resembling cutis verticis gyrata. *Dermatology* 1993;186:294-7.
4. Yazici AC, Ikizoglu G, Baz K, Polat A, Ustunsoy D. Cerebriform intradermal nevus. *Pediatr Dermatol* 2007;24:141-3.
5. Al-Bedaia M, Al-Khenaizan AS. Acromegaly presenting as cutis verticis gyrata. *Int J Dermatol* 2008;47:164.
6. Thappa DM, Sethuraman G, Kumar GR, Elangovan S. Primary pachydermoperiostosis: A case report. *J Dermatol* 2000;27:106-9.
7. Hayashi Y, Tanioka M, Taki R, Sawabe K, Kore-eda S, Utani A, *et al.* Malignant melanoma derived from cerebriform intradermal naevus. *Clin Exp Dermatol* 2009;34:e840-2.
8. Van Raamsdonk CD, Griewank KG, Crosby MB, Garrido MC, Vemula S, Wiesner T, *et al.* Mutations in GNA11 in uveal melanoma. *N Engl J Med* 2010;363:2191-9.
9. Lambert WC, Brodtkin RH. Nodal and subcutaneous cellular blue nevi. A pseudometastasizing pseudomelanoma. *Arch Dermatol* 1984;120:367-70.
10. Rabinovitz HS, Barnhill RL. Benign melanocytic neoplasms. In: Bologna JL, Jorizzo JL, Schaffer JV, editors. *Dermatology*. 3rd ed. USA: Elsevier; 2012. p. 1861.
11. Rompel R, Meyer W, Petres J. Cerebriform intradermal nevus. *Hautarzt* 1993;44:238-41.

Cite this article as: Sarkar S, Roychoudhury S, Shrimal A, Das K. Cerebriform intradermal nevus presenting as cutis verticis gyrata with multiple cellular blue nevus over the body: A rare occurrence. *Indian Dermatol Online J* 2014;5:34-7.

Source of Support: Nil, **Conflict of Interest:** None declared.