Giant cerebriform intradermal nevus in a young girl

Sir,

A 13-year-old girl presented with complaints of a boggy mass on her left frontoparietal scalp since birth with thinning of the overlying hair. The lesion was first noticed at birth and gradually increased in size with age. On examination, a large (12 × 14 cm) soft to firm, bosselated, skin-colored plaque was seen involving the vertex and left frontoparietal scalp. The surface was irregular with convolutions and depressions and there were prominent follicular openings with sparse overlying hair. Solitary (1 × 1 cm), erythematous, pedunculated, firm nodule was noted over the surface [Figure 1]. A punch biopsy from the plaque and an excisional biopsy of the overlying nodule, showed the presence of intradermal nevoid cells in nests with variable amounts of pigmentation. The upper dermal nevoid cells showed dense pigmentation, which was minimal in the deeper dermal nest. These features were suggestive of a cerebriform intradermal nevus [Figure 2]. These nevoid cells stained positive with HMB45 antibody on immunohistochemistry [Figure 3]. The patient was referred to the plastic surgery department for tissue expansion followed by excision.

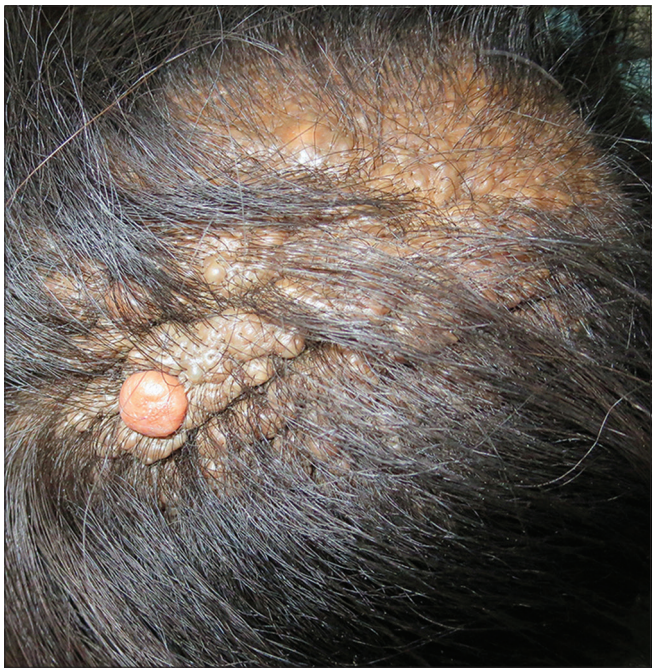

Figure 1: A large (12 × 14 cm) soft, bosselated skin-colored plaque involving the vertex and left frontoparietal scalp

First described by Hammond and Ransom in 1937, cerebriform intradermal nevus is a congenital intradermal nevus that has a brain-like surface contour with bosselations, commonly seen on the scalp. Other sites described are the neck, legs, buttocks, scrotum, and back.[1] It is one of the causes of secondary cutis verticis gyrata, other causes being amyloidosis, syphilis, acromegaly, myxedema, pachydermoperiostosis, pituitary tumors, intracerebral aneurysm, tuberous sclerosis, dermatofibroma, neurofibroma, cutaneous focal mucinosis, and giant congenital melanocytic nevus, which is a close clinical differential diagnosis. Clinically, cerebriform intradermal nevus is a nonpigmented plaque with sparse hairs in contrast to giant melanocytic nevus, which is darkly pigmented and hairy.[2] 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).[1]

Histopathologic examination shows intradermal nevoid cells in nests or lying singly with variable amounts of pigment. There may be neural transformation seen at places, especially in the deeper dermis. Verocay bodies have also been described. The hair follicles are normal or atrophied, and the dermoepidermal junction is spared. These features are in contrast to giant congenital melanocytic nevus, which shows dermoepidermal activity, increased hair follicles and high melanin content.[2]

These nevi have a 4%–10% risk of developing malignant melanoma. Development of firm nodule, pain, ulceration, and a sudden increase in the size of plaque should lead to the suspicion of malignancy, and histopathological evaluation should be done.[3] The risk of neurocutaneous melanosis in patients with cerebriform intradermal nevus has not yet been established.[1] Association with blue rubber bleb nevi is reported.[4] Isolated nevus and the nevus associated with the syndrome are clinically indistinguishable. Surgical excision is

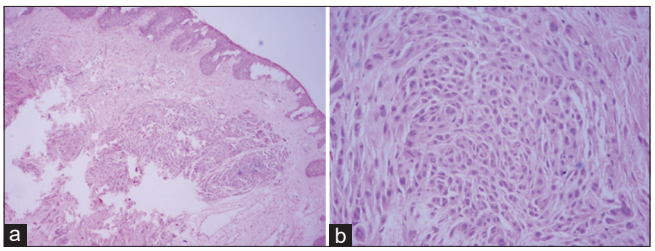

Figure 2: Hematoxylin and eosin stain of skin biopsy. (a) (×10) shows nevoid collection of cells in upper and mid-dermis with pigment in the upper dermal collection. (b) (×40) shows these collections are composed of monomorphic large nevoid melanocytes with vesicular nuclei and abundant cytoplasm

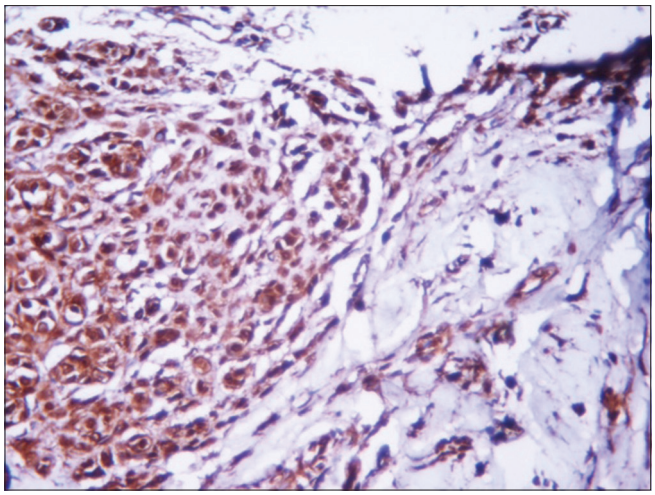

Figure 3: HMB45 stain (×40) of skin biopsy: Nevoid cells shows positive stain

the management of choice, with a postsurgical recurrence risk of 3%–4%.[5]

Thus, these lesions require close follow-up and complete excision whenever feasible due to the high risk of developing malignant melanoma. Scalp reconstruction using complex flaps is often required.

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Conflicts of interest

There are no conflicts of interest.

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REFERENCES

1. Yazici AC, Ikizoglu G, Baz K, Polat A, Ustunsoy D. Cerebriform intradermal nevus. Pediatr Dermatol 2007;24:141-3.
2. Tagore KR, Ramineni AK. A case of cutis verticis gyrata secondary to giant cerebriform intradermal nevus. Indian J Pathol Microbiol 2011;54:624-5.
3. 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.
4. 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.
5. Goodson AG, Florell SR, Boucher KM, Grossman D. Low rates of clinical recurrence following biopsy of benign to moderately atypical dysplastic melanocytic nevi. J Am Acad Dermatol 2010;62:591-6.

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