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Case report

Giant congenital melanocytic nevus with neurofibroma-like lesions and onset of vitiligo

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Abstract

Giant congenital pigmented nevus and neurofibromatosis type 1 may rarely occur together. We reported here an unusual case where giant congenital melanocytic nevus was associated with neurofibroma-like lesions and vitiligo, emphasizing the clinical and histological diagnostic difficulties posed by this presentation, the signification of vitiligo which can testify of a possible malignant transformation of the giant nevus to a melanoma, and highlights the importance of an accurate diagnosis and a close follow-up of such patients.
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Keywords: Giant congenital melanocytic nevus; Neurofibromatosis; Melanoma; Vitiligo; Neurotization

1. Introduction

Congenital melanocytic nevus (CMN) is a benign neoplasm composed of nevomelanocytes. Giant congenital melanocytic nevus (GCMN) is a rare variety of CMN characterized by its size (Bhagwat et al., 2009), and its potential for transformation into malignant melanoma, it is infrequently associated with other findings which make the clinical picture complex. We report here a rare association of GCMN, neurofibroma-like lesions and vitiligo (Gulati et al., 2000).

2. Case report

A 39 year old lady, presented at birth a confluent area of pigmentation covering the trunk, and numerous other pigmented lesions over the limbs. In early adolescence she developed soft, pedunculated brown-colored nodules that were localized mainly on the back and left shoulder. Two years before presenting to us she developed white macular lesions on the hands, foot, face, and also in the area of the GCMN. No other family member was similarly affected. Clinical examination revealed a confluent area of pigmentation over the trunk (back, chest, shoulders, and neck) covered with long hairs consistent with a giant bathing trunk nevus, with pachydermatous changes by location (Fig. 1). In addition numerous other pigmented nevi over the limbs and face were noted (Figs. 2 and 3). There were multiple nodules present over the back, the largest being about 20 cm in diameter; the nodules were soft, with a smooth surface, non pulsatile, non tender and freely mobile on palpation (Fig. 4). Dermoscopic examination found a homogeneous hyperpigmentation, perifollicular hypopig-

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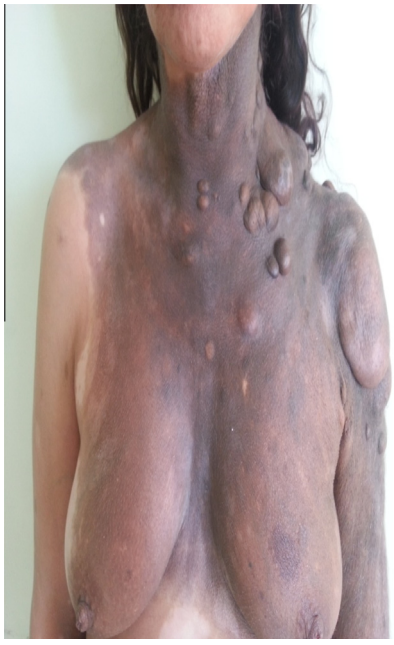

Figure 1. Confluent pigmentation over the trunk with pachydermatous changes by location.

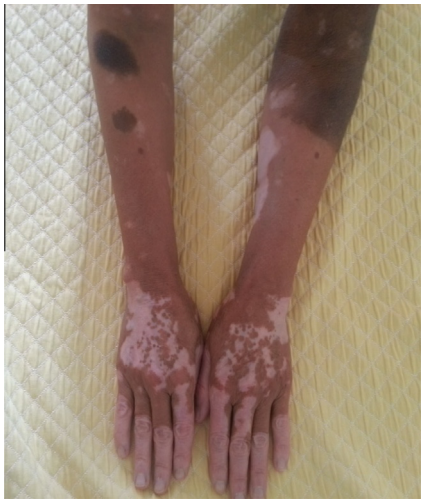

Figure 2. Multiple melanocytic nevi of variable sizes on upper and lower limbs with lesions of vitiligo.

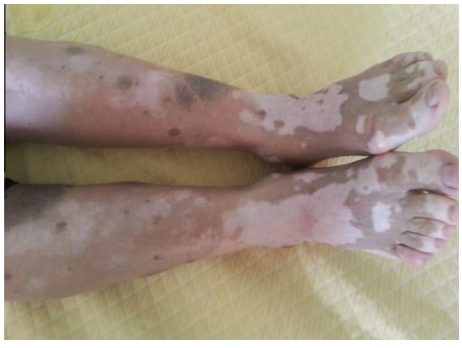

Figure 3. Multiple melanocytic nevi of variable sizes on upper and lower limbs with lesions of vitiligo.

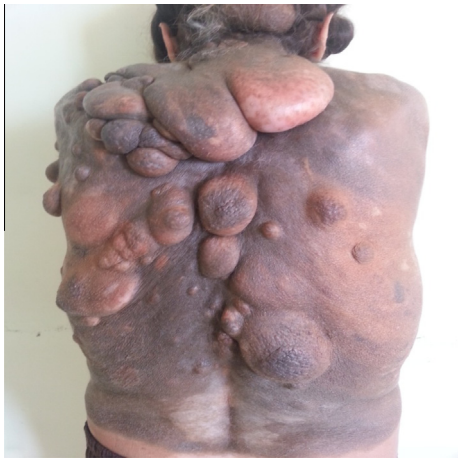

Figure 4. Bathing trunk melanocytic nevus with large, pendulous skin lesions mimicking neurofibromas over the trunk (back, chest, shoulders).

mentation and terminal hairs, in favor of a congenital nevus.

The occipital area was covered by convoluted folds of thickened pigmented skin resembling cutis verticis gyrata associated with a localized alopecia (Fig. 5). Also, multiple white macules were distributed over the area of NCG, face, and lower and upper extremities (Figs. 2 and 3).

There were no cutaneous features to support the diagnosis of neurofibromatosis type 1, and in particular she had no axillary freckling. Ophthalmological assessment, especially examining for the presence of Lisch nodules, and neurological examination were both normal. The biological assessment found hyperthyroidism with anti-thyroid peroxidase antibodies, anti-thyroglobulin and anti TSH-receptor antibodies. Skeletal X-ray was normal, and brain and spinal magnetic resonance imaging (MRI) in search of neurocutaneous melanocytosis and spina bifida occulta is not effected due to the lack of means. Biopsies performed at

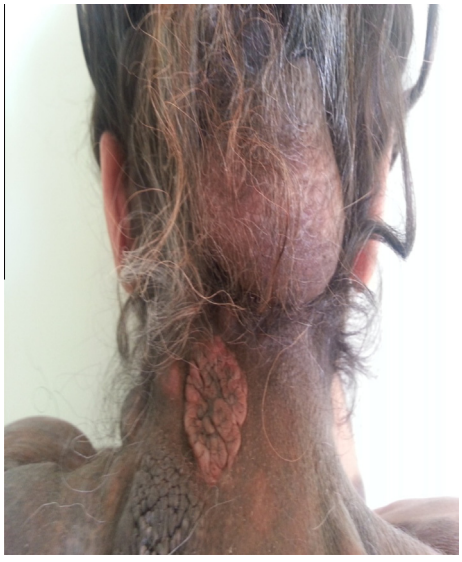

Figure 5. Convoluted and pigmented folds of skin in the occipital area of the scalp with associated alopecia.

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