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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.

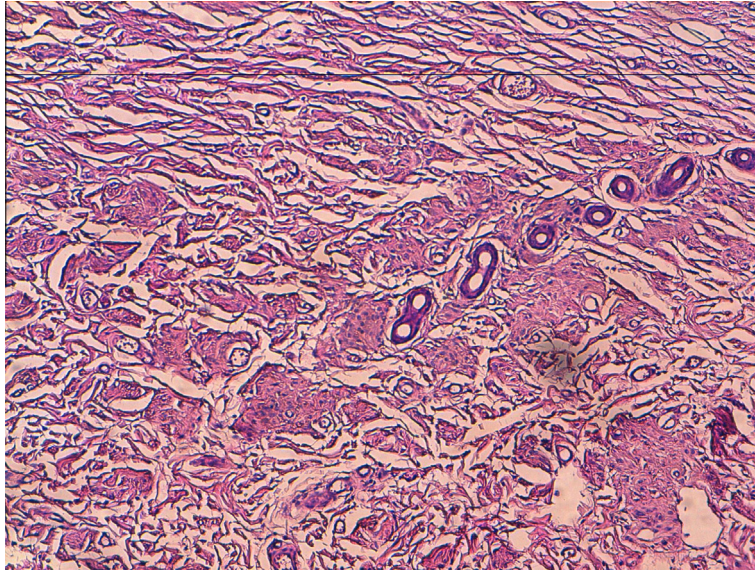


Figure 6. Haematoxylin and eosin, $\times 20$. Melanocytes become spindle shaped within the reticular dermis.

multiple sites were all in favor of a congenital nevus which has undergone marked neurotization on the nodules (Fig. 6).

3. Discussion

We report an original case of GCMN associated with neurofibroma-like lesions and vitiligo.

GCMN also known as Bathing trunk nevus (BTN) or garment nevus, is fortunately uncommon and has been arbitrarily defined by size (lesion greater than 20 cm diameter), or as a lesion that occupies a region considered major such as the face, or one that cannot be excised without resulting in significant deformity. It presents at birth in 1 in 500,000 newborns with a female predominance, like a large dark-brown plaque with excessive growth of hair, and grows in proportion to the site of the body on which it is located (Sasmaz et al., 2005). The mode of inheritance is probably multi factorial. 82% of the GCMNs occur in an axial distribution (trunk, head, and/or neck), with a possible association to spina bifida, meningocele, vascular nevi, lipomas, Dandy–Walker malformation, arachnoid cysts, and Chiari type 1 malformation (Ansarin et al., 2006). Satellite nevi may be present in 74–91% of the GMNs. They can also be associated with nevi in the central nervous system as a distinctive syndrome: neurocutaneous melanocytosis (NCM) or nevomatosis, in our case the neurological examination and the brain and spinal MRI were normal.

There is a very rare form of GCMNs which can mimic neurofibromatosis type I, but GCMN, has been also reported to occur in about 5% of patients with von Recklinghausen's neurofibromatosis (Ball and Kho, 2005). In our patient, the diagnosis of neurofibromatosis type I was eliminated because no other clinical diagnostic criteria

were present. Heuer (1917) first drew attention to the relationship between bathing trunk nevi and Von Recklinghausen's fibromatosis, he reported 26 cases of GCNMC resembling NF type 1, including skin pigmentation, fibromas or other cutaneous tumors and neurofibromas.

Reed et al. (1965) recognized the clinical and occasional microscopic similarity between giant congenital hairy nevi and NF1 but they considered these to be totally separate entities (Silfen et al., 2002).

Therefore, not all neurofibroma-like lesions detected clinically in cases with BTN are real neurofibromas (Sasmaz et al., 2005), as it was the case of our patient.

The GCMN has a potential to transformation into so many malignancies, especially malignant melanoma.

People with GCMN have an increased risk of developing neural crest malignancies; most commonly malignant melanoma in at least 6.3%, and in more than 50% of cases, this malignant change occurs prior to puberty (Ansarin et al., 2006); for this reason a policy of early excision of giant bathing trunk nevi has been advocated if possible (Whittam et al., 1996).

Our patient developed vitiligo 2 years ago; vitiligo is an immunologic disease, that is supported by discoveries of antimelanocyte antibodies, vitiligo antigens, and frequent association with other autoimmune diseases, especially in females, bilateral, and acrofacial types of vitiligo, but there are many reports of melanoma associated with vitiligo, and some cases have been related to regression of melanoma or development of metastasis (Shin et al., 2002). That was the second dilemma in our patient; what is the signification of vitiligo? Can we consider it a simple autoimmune pathology based on the following arguments: female sex, acrofacial distribution and the presence of the thyroiditis, or is it a sign of melanoma?

Our patient underwent a dermoscopic examination with biopsies in multiples sites, all were in favor of nevi, however close follow up remains necessary.

Serial excision is the treatment of choice of GCMN, and laser should only be regarded as a treatment option for lesions that cannot be surgically excised. In cases in which surgical excision is not feasible as in our patient, close clinical and dermoscopic follow up with biopsy of clinically suspicious areas is recommended (Ansarin et al., 2006).

This clinical and histopathological similarity between the CMN and neurofibromas is likely to result from the fact that both melanocytes and Schwann cell originate from the neural crest (Silfen et al., 2002).

The term neurotization refers to the resemblance of some melanocytic cells to peripheral nerve sheath cells as we found in biopsy of our patient. Melanocytic cells located in the epidermis and upper dermis, epithelioid or so-called type A cells, have a plump, rounded appearance. In the deeper dermal component of the nevus, cells may have a spindle-shaped appearance and resemble peripheral nerve sheath cells, these have been termed type C cells. Nevus cells in the mid-dermis, which are small and round in appearance, are termed B cells (Whittam et al., 1996). It has been suggested that immunohistochemical markers can be used to differentiate between neurofibromas and neuroid nevi reliably. Nevus cells of all types will stain with S-100 but not with Leu-7 (CD57), myelin basic protein (MBP) or glial fibrillary acid protein (GFAP), whereas neurofibromas stain with Leu-7, MBP and GFAP as well as with S-100 (Gach et al., 2004).

4. Conclusion

Clinicians should be conscious that not all lesions resembling neurofibromas occurring on GCMN are real neurofibromas. The risk of melanoma in such patients imposes close clinical and histological follow-up, especially if there is an added vitiligo.

Conflict of interest

None.

References

- Ansarin, H., Soltani-Arabshahi, R., Mehregan, D., Shayanfar, N., Soltanzadeh, P., 2006. Giant congenital melanocytic nevus with neurofibroma-like changes and spina bifida occulta. *Int. J. Dermatol.* 45 (11), 1347–1350.
- Ball, N.J., Kho, G.T., 2005. Melanocytic nevi are associated with neurofibromas in neurofibromatosis, type I, but not sporadic neurofibromas: a study of 226 cases. *J. Cutan. Pathol.* 32 (8), 523–532.
- Bhagwat, P.V., Tophakhane, R.S., Shashikumar, B.M., Noronha, T.M., Naidu, V., 2009. Giant congenital melanocytic nevus (bathing trunk nevus) associated with lipoma and neurofibroma: report of two cases. *Indian J. Dermatol. Venereol. Leprol.* 75 (5), 495–498. <http://dx.doi.org/10.4103/0378-6323.55394>.
- Gach, J.E., Carr, R.A., Charles-Holmes, R., Harris, S., 2004. Multiple congenital melanocytic naevi presenting with neurofibroma-like lesions complicated by malignant melanoma. *Clin. Exp. Dermatol.* 29 (5), 473–476.
- Gulati, R., Jain, D., Mehraania, K., Kuldeep, C.M., Mathur, D., 2000. Giant congenital nevocmelanocytic nevus with satellite lesions, vitiligo and lipoma: a rare association. *Indian J. Dermatol. Venereol. Leprol.* 66 (6), 316–317.
- Heuer, S., 1917. Ein Fall von ausgedehntem schimmohsenartigem naevus pigmentosus pilosus congenitus mit Hamartom des Ruckens und spina bifida occulta: seine beziehung zur V Recklinghausen's Schen Krankheit. *Bruns Beitr Klin Chir* 104, 388.
- Reed, W.B., Becker, S.W., Becker Jr., S.W., 1965. Giant pigmented, melanoma, and leptomenigeal melanocytosis: a clinical and histopathological study. *Arch. Dermatol.* 91, 100.
- Sasmaz, S., Celik, S., Ciralik, H., Arican, O., Gumusalan, Y., 2005. A case of bathing trunk nevus studded with neurofibroma-like papules. *Clin. Dysmorphol.* 14 (1), 41–43.
- Shin, J.H., Kim, M.J., Cho, S., Whang, K.K., Hahm, J.H., 2002. A case of giant congenital nevocytic nevus with neurotization and onset of vitiligo. *J. Eur. Acad. Dermatol. Venereol.* 16 (4), 384–386.
- Silfen, R., Skoll, P.J., Hudson, D.A., 2002. Congenital giant hairy nevi and neurofibromatosis: the significance of their common origin. *Plast. Reconstr. Surg.* 110 (5), 1364–1365.
- Whittam, L.R., Macdonald, D.M., Hay, R.J., 1996. A case of giant bathing trunk naevus with neurofibroma-like change. *Clin. Exp. Dermatol.* 21 (2), 167–169.