Congenital hairy melanocytic nevus associated with vitiligo

Taghreed Jameel Almaaita, Salah Abdallat

Department of Dermatology, Royal Medical Services, Amman, Jordan

ABSTRACT

We report here an association between giant hairy melanocytic nevus and vitiligo in a 7 year old male presented with giant congenital hairy melanocytic nevus since birth with no other medical illnesses. It affected about 80% of back and for the last 3 years it extended over his face. (Rawal Med J 2014;39: 240-241).

Key words: Vitiligo, melanocytic nevus.

INTRODUCTION

Congenital melanocytic nevi (CMN) are nevi that are present at birth.Small lesions are most often unapparent but large nevi can carry a psychosocial burden and increased risks of malignant melanoma (MM).[1] Exact definition for Giant congenital melanocytic nevus (GCMN) with some authors includes a surface area of 20cm,[1-3] while others have used varying body surface area measurements or other definitions.[4-6] We will use the term GCMN to nevi measuring 20 cm. Regardless of the size it can be associated with MM, and Neurocutaneous melanosis.[1]

Vitiligo is an acquired disorder of the skin and mucous membranes,[6] and may appear at any age.[7] Approximately 0.5% to 1% of the population is affected and ratio appears to be equal between men and women.[7] It can be a psychologically burden especially in darker skinned individuals, in whom it is more noticeable. The actual pathogenesis is under debate and has been attributed to autoimmune causes.[6] Vitiligo can be divided into two major classes: non segmentalvitiligo (NSV), which is more common and segmentalvitiligo (SV).We present here a distinguished association between giant hairy melanocytic nevus and vitiligo in a patient which is not present in any case in literature.

CASE PRESENTATION

A 7 year old male presented to dermatology clinic with giant melanocytic nevus since birth. It was completely asymptomatic except for the disfiguring appearance. He was investigated thoroughly after birth but no other birth defect was found and there was no other complaint. It affected about 80% of back with some extension to the side of the abdomen (Fig 1). Over the last 3 years he developed depigmented vitiligenous lesions over his face mainly perioricular and over the forehead. Full clinical history was completely normal. No significant family history of similar problems or malignant melanoma was found.

Clinical examination revealed the presence of large melanocytic lesion involving about 80% of his back and extended anteriorly to the sides of the abdomen. No associated lymph node enlargement was seen. There was perioricular depigmented lesion and similar lesion on the forehead. The facial depigmentedarea was examined by Woods light and it was milky white suggestive of vitiligo.

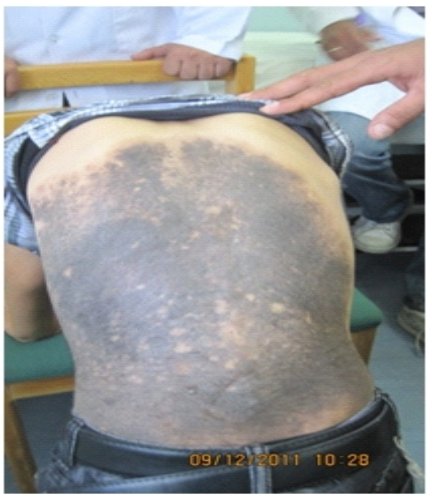

Fig 1. Congenital hairy melanocytic nevus.

Three skin biopsies were done from different part of the giant nevus and it showed melanocytic proliferation with nesting pattern superficially and extended to the deep dermis and deep adnexal areas as hair follicle with minor extension to

subcutaneous tissue which proved the diagnosis of GCMN and no melanoma was found. Skeletal and brain MRI were normal and skull x ray had frontal bossing but it was not of clinical significance,

DISCUSSION

Vitiligo typically occurs in uncovered areas and has a major impact on self-esteem. In some societies, women with vitiligo have difficulty to engage[8] or study.[9] Many worry about the disease worsening, have their social life affected and feel depression. Exacerbated factors include severe sunburn,[10] pregnancy,[10,11] skin trauma[11] and/or emotional stress.[10,11] A significantly higher incidence of koebnerization and disease progression is seen in NSV.

Nevus cells are derived from neural crest melanocytes.[12] Many CMN have been found to harbor N-Ras mutations.[13,14] This differs from acquired nevi and melanomas arising on intermittently sun-exposed skin, which typically have B-RAF mutations.[14] In addition, some suggest a genotype phenotype correlation for CMN size and mutation types.[13] Specific dermatoscopic features for CMN include argot network, globules, and perifollicular hypopigmentation.[1] Skin biopsy findings that support the diagnosis include a presence of deep nevus cells, particularly reaches adnexal structures.[1]

We found different association between vitiligo and many condition rare or common especially with other endocrine and autoimmune disorder. And the same is about congenital melanocytic nevus as it is associated with a wide spectrum of other congenital defect but the feature in our patient was the unique association between GCMN and vitiligo, which is the first in the literature.

Corresponding author email:anoud1998@yahoo.com
Conflict of Interest: None declared
Rec. Date: Jan 31, 2013 Accept Date: Mar 19, 2013

REFERENCES

1. Alikhan A, Ibrahimi OA, Eisen DB. Congenital melanocytic nevi: Where are we now? Part I. Clinical presentation, epidemiology, pathogenesis, histology malignant transformation and neurocutaneousmelanosis. J Am Acad Dermatol 2012;67: 495.e1-17.
2. Mark GJ, Mihm MC, Liteplo MG, Reed RJ, Clark WH. Congenitalmelanocytic nevi of the small and garment type. Clinical,histologic, and ultrastructural studies. Hum Pathol 1973;4:395-418.
3. Quaba AA, Wallace AF. The incidence of malignant melanoma(0 to 15 years of age) arising in "large" congenitalnevocellular nevi. Plast Reconstr Surg 1986; 78:174-81.
4. Ruiz-Maldonado R. Measuring congenital melanocytic nevi. Pediatr Dermatol 2004; 21:178-9.
5. Swerdlow AJ, English JS, Qiao Z. The risk of melanoma inpatients with congenital nevi: a cohort study. J Am Acad Dermatol 1995;32:595-9.
6. Kyriakis KP, Palamaras I, Tsele E, Michailides C, Terzoudi S. Case detection rates of vitiligo by gender and age. Int J Dermatol 2009;48:328-9.
7. Taieb A, Picardo M. Clinical practice. Vitiligo. N Engl J Med 2009; 360:160-9.
8. Zhang Z, Xu SX, Zhang FY, Yin XY, Yang S, Xiao FL, et al. The analysis of genetics and associated autoimmune diseases in Chinese vitiligo patients. Arch Dermatol Res 2009; 301:167-73.
9. Porter JR, Beuf AH. Racial variation in reaction to physicalstigma: a study of degree of disturbance by vitiligo amongblack and white patients. J Health Soc Behav 1991;32:192-204.
10. Manolache L, Benea V. Stress in patients with alopecia areataand vitiligo. J Eur Acad Dermatol Venereol 2007; 21:921-8.
11. Baris-Drusko V, Rucevi I. Trigger factors in childhoodpsoriasis and vitiligo. Coll Antropol 2004; 28:277-85.
12. Jiao Z, Zhang ZG, Hornyak TJ, Hozeska A, Zhang RL,Wang Y, et al. Dopachrometautomerase (Dct) regulatesneural progenitor cell proliferation. Dev Biol 2006;296:396-408.
13. Wilkie AL, Jordan SA, Jackson IJ. Neural crest progenitors of the melanocyte lineage: coat colour patterns revisited. Development 2002;129:3349-57.
14. Bauer J, Curtin JA, Pinkel D, Bastian BC. Congenital melanocytic nevi frequently harbor NRAS mutations but no BRA Fmutations. J Invest Dermatol 2007;127:179-82.