Author’s response to reviews

**Title:** An unusual case of congenital melanocytic nevus presenting as neurocutaneous melanoma coexisting with Tuberous Sclerosis complex: a rare association or a new syndrome?

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**Version:** 3  **Date:** 16 March 2011

**Author’s response to reviews:** see over
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Version: 2

Author's response to reviews: see over
Thank you for consideration of our manuscript for publication in your journal.

We have reviewed the above manuscript according to your reviewer’s comments.

Reviewer #1 (Prem Sagar Subramanian)

Comments to authors: (all comments by reviewers are highlighted in yellow)

Comment 1 by reviewer:

1. The authors present an interesting case with association of TSC and NCM; while pigmented lesions can occur in TSC, NCM has not been reported previously. The case is well-presented, but one important question remains about the diagnosis of NCM vs congenital melanocytic nevus. The latter has certainly been reported before, and the lack of brain MRI features of leptomeningeal melanin deposition does call the diagnosis of NCM into question. What specific aspects of this patient’s disease are more consistent with NCM than with CMN? These features are not clear from the text and are critical to the uniqueness of this case.

Answer to comment 1:

Considering the classification/criteria put forth by Kadonaga and Frieden, the diagnosis of NCM cannot be put into question. It is not necessary that cases of NCM should/must compulsorily have MRI features of leptomeningeal deposition. Kindly see below the criteria put forth by Kadonaga and Frieden in 1991 for diagnosis of NCM and our patient fits in the criteria 3 (highlighted in blue).

- In support of NCM (features consistent with NCM):

NCM is currently diagnosed by the following criteria put forth by Kadonaga and Frieden in 1991: (1) large and/or multiple CMN in association with meningeal melanosis or melanoma; (2) no evidence of cutaneous melanoma, except in patients with histologically benign meningeal lesions; or (3) no evidence of meningeal melanoma, except in patients with histologically benign cutaneous lesions.

Reference from text/manuscript:

Our patient had a single large-sized CMN with an axial distribution over the scalp with no evidence of meningeal melanoma but had histologically benign cutaneous scalp lesions pointing towards the diagnosis of NCM; thus meeting the criteria outlined in point number (3) as proposed by Kadonaga and Frieden in 1991, which is now the most acceptable criteria used for the diagnosis of NCM rather than just the presence of MRI Brain features of leptomeningeal melanin deposition.

- **In support of CMN (Features consistent with CMN):**

  Congenital Melanocytic Nevus (CMN) are one of several known risk factors for the eventual development of melanoma. CMN are classified into large (≥ 20 cm), medium (1.5 to 19.9 cm) and small (≤1.5cm) nevi according to their size [9]. The term "multiple" is used when more than three lesions are present.

  **Reference from text/manuscript:**
  

  Our patient was born with a triangular scalp nevus covered with hair. She also had multiple black hairy patches on her extremities, back and most of the anterior trunk. The scalp nevus however grew over a period of 8 years from birth to the present massive size and thus presenting as a melanoma. Also the histological findings were consistent with CMN. It is this CMN which is now presenting as a melanoma. Hence the title, Congenital melanocytic nevi presenting as Neurocutaneous melanoma (NCM) was chosen.

**Comment 2 by reviewer:**

2. Quality of written English: Acceptable

- Thank you
The article describes a rare association, which merits publication. However, the manuscript has a few shortcomings that need to be addressed:

Comment 1 by reviewer:
1. The grammar and language usage leaves much to be desired. Kindly rewrite the manuscript, especially the case-description under the guidance of a good English user/Senior colleague.
   • Done.
   • All the grammatical errors and the corrections advised by the reviewers have been made and included in the manuscript. The additions made in the manuscript have also been highlighted in the manuscript.

Comment 2 by reviewer:
2. Some major flaws in description or writing have been highlighted and suggestions provided in the margins.
   • Done

Comment 3 by reviewer:
3. ‘Neurocutaneous melanoma coexisting with Tuberous Sclerosis complex’ could be a more apt title.
   • Our patient was born with a triangular scalp nevus covered with hair. She also had multiple black hairy patches on her extremities, back and most of the anterior trunk. The scalp nevus however grew over a period of 8 years from birth to the present massive size and thus presenting as a melanoma. Also the histological findings were consistent with CMN. It is this CMN which is now presenting as a melanoma. Hence the title, Congenital melanocytic nevi presenting as Neurocutaneous melanoma (NCM) was chosen. Tuberous sclerosis was just a coexistent findings.

Comment 4 by reviewer:
4. Age of the patient at the time of writing the manuscript is not clear. Was the first presentation at 11 years and she was followed for 8 years OR did she present at 11 years and was operated upon. If the latter, how is it that age of menarche (15 yrs) is mentioned? If the former, kindly make necessary changes in the abstract, introduction and case-report.
The confusion was due to a typographical error. The age of the patient at the time of presentation at our institute was 16 years and not 11 as mentioned earlier. This was a typographical error and we apologize for the confusion. She attained her menarche at the age of 15 years as mentioned in the manuscript. The changes have been made in the manuscript.

Comment 5 by reviewer:
5. You need to reduce the length case –description slightly and make the discussion more focused
   • Done (The discussion has been made more focussed as suggested)

Comment 6 by reviewer:
6. Kindly reduce the size of the abstract.
   • Done (The abstract has been reduced)

Comment 7 by reviewer:
7. Were no plastic surgeons or neurosurgeons involved in the management?
   • The case was operated at the Rural Medical College, Loni, India. No plastic surgeons and neurosurgeons were involved in the operation due to non affordability by the patient and her parents. Excision with primary skin grafting was done. Graft was taken from the medial aspect of the right thigh.

Comment 8 by reviewer:
Quality of written English: Not suitable for publication unless extensively edited
   • Done (Changes made)