Letter in Response to Previous Publication

An unusual nevus spilus and neurofibromatosis type 1

Sir,

A growing wealth of clinical and molecular data supports the view that pigmented lesions such as segmentally arranged lentigines, segmentally arranged café-au-lait macules and giant café-au-lait macules can be considered phenotypic variants of mosaic neurofibromatosis type 1. In a recent issue of the Indian Journal of Dermatology, Venereology and Leprology, Erdem *et al*. reported a case of Lisch nodules and pilocytic astrocytoma associated with large hypermelanotic patches lateralized on the right side of the body and featuring superimposed, scattered maculopapular naevi.[1] Such cutaneous manifestations were labeled by the authors as "partial unilateral lentiginosis," but are instead fully consistent with a diagnosis of papular nevus spilus. The histological features of lentigo simplex do not constitute a compelling argument since such pattern can very well be found in the macular speckles of papular nevus spilus.

There are only scant published data concerning the frequency of the occurrence of nevus spilus of any type (small/segmental, macular/papular) in patients with neurofibromatosis type 1, but it might not differ from that in the general population.[2] In particular, reports of patients affected by a segmental papular nevus spilus and stigmata of neurofibromatosis type 1 are extremely rare.[3-5] Intriguingly, these cases are similar to the one by Erdem *et al*.[1] in that: (1) they displayed a mild phenotype which always included Lisch nodules (often mentioned as being ipsilateral to the nevus); and (2) papular nevus spilus was always characterized by very small and quite sparse maculopapular nevi. It could be hypothesized that these lesions do not represent true papular nevus spilus but rather neurofibromatosis type 1-related, segmental café-au-lait macules. However, café-au-lait macules do not specifically harbor melanocytic nevi, which indeed are not known to be caused by neurofibromatosis type 1 gene mutations. On the other side, it is unlikely that the above-mentioned papular nevus spilus cases are associated with neurofibromatosis type 1 by mere chance, if only considering the homolaterality of most clinical manifestations. Increased reporting and molecular studies will hopefully help shedding light on the seemingly rare co-occurrence of such "paucipapular" nevus spilus and neurofibromatosis type 1 stigmata.

Declaration of patient consent

Patient's consent not required as there are no patients in this study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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