

Image Letter

Co-localization of nevus depigmentosus with café au lait macules: Pigmentary twin spotting?

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Case Presentation

A 21-year-old woman presented with complaints of hypopigmented lesions over right side of her face since infancy along with gradually progressive hyperpigmented lesions over the same side since nearly 15 years of age. On examination, multiple discreet to confluent, irregular, hypopigmented macules were present over right side of the face, overlying which, multiple irregularly shaped hyperpigmented macules were also noted along with few scattered freckles (Figure 1A). Dermoscopy (Heine Delta 20T, 10x) of hypopigmented lesions revealed white structureless areas (Figure 1B) and from hyperpigmented lesions showed dark brown reticular pattern of pigmentation with brown dots (Figure 1C).

The patient did not reveal any similar history in other family members. Systemic examination was non-significant. Hence a final diagnosis of twin spotting of nevus depigmentosus with café au lait macule was made.

Teaching points

Naevus depigmentosus is a disorder of hypomelanosis through postzygotic somatic mutations, resulting in reduced melanogenic potential in a clone of cells during embryogenesis [1]. Café au lait macules (CALMs) are brown coloured maculae with increased melanin and melanocytes within the epidermis. They can be seen sporadically and in association with several genetic syndromes such as neurofibromatosis 1 and Legius syndrome [1].

Twin spotting refers to the presence of adjacent clones of genetically different cells in a background of normal cells producing different phenotypes. It has been hypothesized that these lesions have separate recessive genetic mutations occurring on the same chromosome [2].

Hereby we report an interesting and rare presentation of CALMs and Naevus depigmentosus co-localizing on right side of the face which suggests twin spotting.

References

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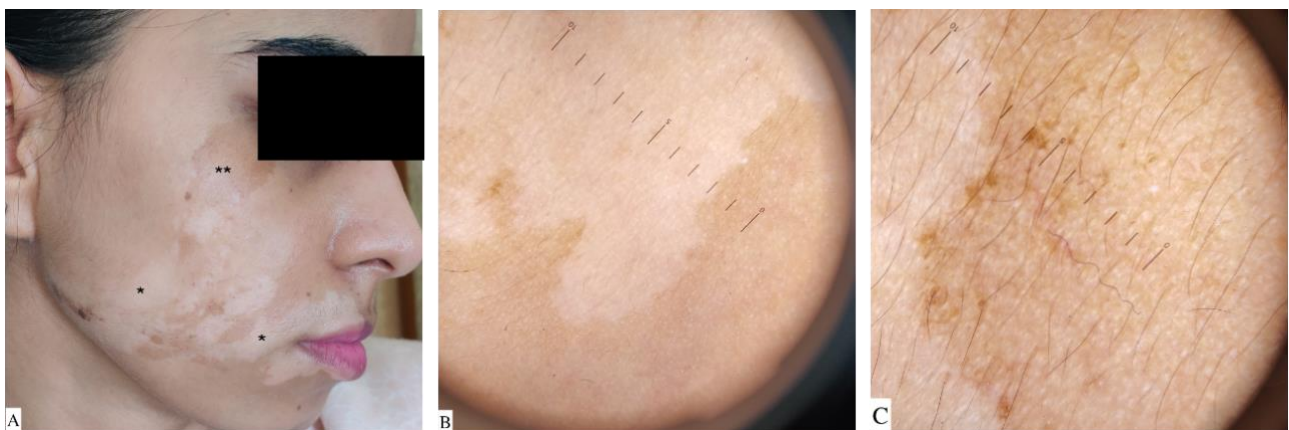


Figure 1. A) Hyperpigmented (**) macules suggestive of CALMs and hypopigmented macules(*) suggestive of naevus depigmentosus over ipsilateral side of the face. B) and C) Dermoscopy from naevus depigmentosus and CALM showing white structureless areas and dark brown reticular pattern of pigmentation with brown dots respectively (Heine Delta 20T, 10x).