Case 24

A 20-year-old Thai female from Bangkok

Chief complaint: Asymptomatic hypo- and hyperpigmented patches on back, right shoulder, right arm & right upper chest since birth

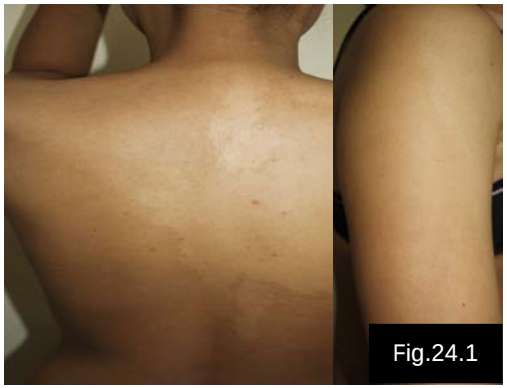

Fig.24.1

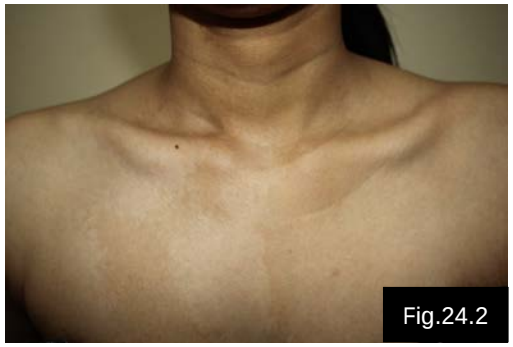

Fig.24.2

Present illness:

She developed multiple, asymptomatic, hypo- and hyperpigmented patches on back, right shoulder, right arm and right upper chest since birth. The pigmented lesions gradually increased in size along with her growth. She was ortherwise healthy.

Past history: No underlying disease, normal growth and development

Family history: No other family member had similar lesion

Physical examination: Unremarkable

Dermatologic examination:

- Well circumscribed hyper- and hypopigmented patches in close proximity to each other in a background of normal skin on back, right upper chest, shoulder and arm (Fig.24.1, 24.2)

Histopathology: Not done

Investigation: Not done

Diagnosis: Cutis tricolor

Treatment: Reassurement

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Discussion:

Cutis tricolor is a skin abnormality consisting in a combination of congenital hyper- and hypopigmented skin lesions (in the form of paired macules, patches or streaks) in close proximity to each other

in a background of normal skin.[1-4] This is a rare disease which reflects a mechanism called twin-spotting phenomenon. It is postulated that the underlying gene locus of this phenomenon is a hot spot for post zygotic recombination resulting in two different mutant alleles located on the same locus of homologous chromosome and clinically explaining the coexistence of three different complexions. The earlier the mutation occurs during embryogenesis, the wider the involvement of the skin, skeletal, CNS and other structures will be. Conversely, later post zygotic mutation causes only skin involvement. This event is usually sporadic, however, familial occurrence has been reported in two sisters.[3,5,6]

The lesions usually developed at birth or in the first months of life.[2,4] The cutaneous lesions present as macules, patches and streaks which arrange in different typical (e.g., patchy, linear, sash-like, diffuse) patterns that involve large body areas.[1,3,4] The degree and the extent of the pigmentary anomaly can increase in the first year of life then they stabilize and could also fade in some areas.[1] This phenomenon has been reported in 4 types[5]: (I) an purely cutaneous trait [7-10]; (II) a part of a complex malformation phenotype (Ruggieri-Happle syndrome, RHS) which has facial (a consisting in coarse, asymmetric dolichocephaly, thick and brushy eyebrows, hypertelorism, deep nasal bridge with large bulbous nose and anteverted nostrils, low-set ears, large phyltrum), eye (congenital cataract), skeletal (small skull with prognathism, dystrophic vertebrae and mildly bowed long bones), nervous system (corpus callosum anomalies, white matter abnormalities, holoprosencephaly and cerebellar anomalies) and systemic abnormalities.[11-13]; (III) a distinct type with multiple, disseminated smaller skin macules (cutis tricolor parvimaculata)[6] and different from the autosomal dominant congenital hypo- and hyperpigmented macules (Westerhof syndrome); and (IV) in association with other skin disturbances (e.g. cutis marmorata telangectasica congenital/phacomatosis achromico-melano-marmorata) or in the context of other skin disorders (e.g. ataxia-telangiectasia and phacomatosis pigmentovascularis/PPV) or in neurocutaneous phenotypes.[10,14,15]

The histopathological finding show increase in melanin content of the basal layer but no dermal melanin or melanophages in the hyperpigmented areas and decrease in the melanin content and in the number of melanocytes in the hypopigmented lesions.[16]

For the treatment, skin abnormalities are usually stable over time and do not benefit in laser therapy and extracutaneous involvement such as: skeletal defects are usually mild to moderate & do not progress, the neurological/behavioural phenotype tend to stabilize over time but need for long term investigated follow-up.[1]

In summary, this case is a Thai girl presented with asymptomatic hypo- and hyperpigmented patches on the large areas of the body since birth which typically compatible with cutis tricolor without extracutaneous involvement in the pattern of purely cutaneous trait. Also she was reassured and advised about clinical course and prognosis of the disease and there is no necessary treatment in this case.

References:

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