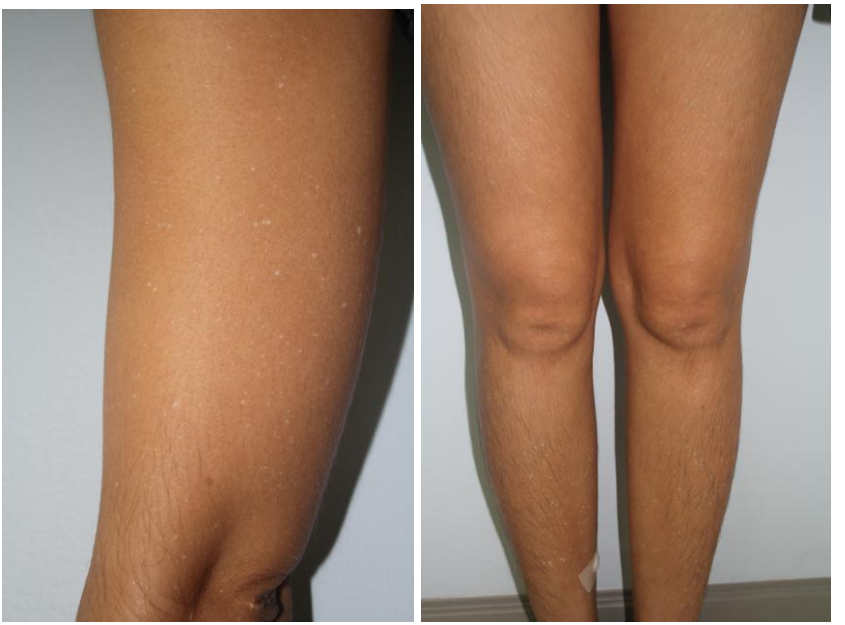
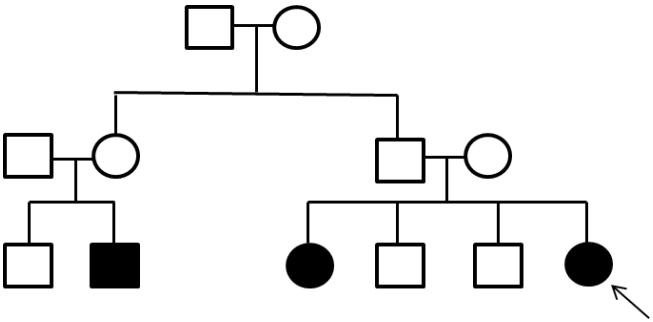
Case 11
A 33-year-old Thai woman from Bangkok.
Chief complaint: Multiple white spots on extremities for 15 years.



Present illness: The patient has developed asymptomatic multiple small hypopigmented and depigmented macules on upper and lower extremities since she was 8 year-old. She was previously diagnosed and treated as vitiligo without any improvement. During the past 2 years, lesions gradually increased in number, especially at exposed area. She denies previous history of frequent sun-exposure and any systemic symptoms.
Past history : No known underlying disease.
Family history: There are 2 family members who have similar dyspigmentation as shown in family pedigree.



Skin examination

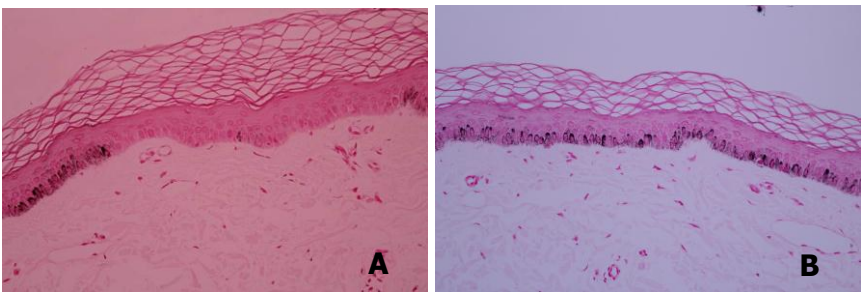
Multiple discrete well-defined hypopigmented and depigmented macules of sizes varying from 2-4 mm on upper and lower extremities predominated at sun exposed area.

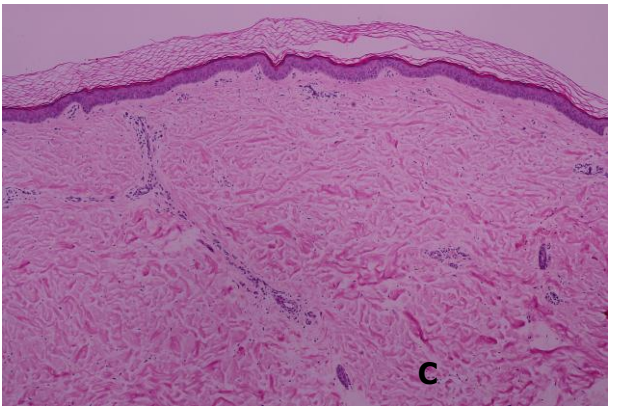
Wood's lamp examination: enhancement of hypo- and depigmentation.

Histopathology

(S13-1130, right lower leg: A. hypopigmented lesion, B. perilesional normal skin)

- Slightly decrease in melanocytes in the epidermis by Fontana–Masson stain (A), comparing with perilesional normal skin (B).
- H&E stain of hypopigmented lesion showed effacement of epidermis with scant epidermal melanin (C).





Diagnosis: Idiopathic guttate hypomelanosis (childhood-onset)
Treatment: Cryotherapy, sun avoidance.
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Discussion

Idiopathic guttate hypomelanosis (IGH) is a common acquired leukoderma characterized by multiple, discrete, round or oval, porcelain-white macules with 2–5 mm in diameter.[1] It usually locates in sun-exposed areas of the skin, especially on the extensor surfaces of arms and legs, but sometimes become generalized.[1] According to a recent study[1], the most commonly involved site is the distal part of the lower extremities, followed by the distal part of the upper extremities and 6% of subjects develop IGH on the face. The prevalence of IGH increases with age and IGH affects up to 87% of adults aged more than 40 years.[1] However, there is a cross-sectional study reports 29.8% of patients have initial lesions before the age of 20 years and found the earliest onset at 3 years of age.[2] IGH is found almost equally in both sexs, but occur earlier in young women than in young men.[1] Hairs within the lesions are not depigmented.[2]

The etiopathogenesis of IGH is obscure. A multifactorial etiology rather than a single cause is likely. Chronic actinic exposure is one of the debated causative factors. Although lesions develop mostly in exposed area of extremities and are observed following narrowband UVB and PUVA therapy[3], but a cause-effect relationship between chronic actinic exposure and the development of IGH could not be establish by statistical studies.[4] A familial tendency to develop this condition has been noted.[5-7] Falabella *et al.*[6] revealed a highly statistically significant preponderance of manifestations of IGH among the relatives of IGH subjects compared with those of normal controls, which clearly indicated a family aggregation among subjects with IGH. Arrunategui *et al.*[7] proposed that the presence of HLA-DQ3 was associated with IGH. Other factors such as trauma, e.g. chronic scrubbing has been also suggested as a causative factors.[1]

The histological findings associate with IGH are hyperkeratosis, atrophic epidermis, flattened rete ridges, and solar elastosis. There is decrease in melanin content, numbers of melanocytes and melanogenic activity.[2] Electron microscopy shows decreased melanosomes, attenuated dendrites, dilatation of the endoplasmic reticulum and swelling of the mitochondria which suggested melanocyte degeneration.[2]

The treatment of choice is still controversial.There are several reports of cryotherapy in treatment of IGH.[8-10] A light 3-5 second liquid nitrogen freeze appears to be sufficient to cause repigmentation.[11, 12] However, the exact mechanism of IGH repigmentation through freezing has not been clearly determined, but freezing time during cryotherapy could probably inactivate an inhibitory enzymes or chemokines related to melanogenesis.[13] Another surgical method is superficial dermabrasion.[8] Fractional carbon dioxide lasers in IGH have been reported as effective and safe treatment option for IGH patient.[9, 10] For medical treatment , topical and intralesional corticosteroid, retinoids, topical tretinoin

and topical calcineurin inhibitor appear to be effective treatments for IGH.[10, 14, 15]

Our patient was diagnosed as IGH according to her clinical presentation and histological findings. Although the early age of onset of IGH in this patient is not very common, but it is possible especially in case of positive family history. After diagnosis, she was instructed to avoid excessive sun exposure and targeted cryospray was initially treated at her right forearm.

References

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