Idiopathic Guttate Hypomelanosis

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Abstract

Idiopathic guttate hypomelanosis (IGH) is characterized by asymptomatic, small, sharply demarcated, hypopigmented or depigmented macules. They are more common on the limbs of elderly persons with sundamaged skin. However, they also occur as discrete lesions, even in sunprotected areas of skin, in young adults. Some patients are quite concerned about the cosmetic appearance of these depigmentations. Many treatments have been described with variable success. IGH lesions can be treated with cryotherapy effectively. This is most suitable where there are only a small number of lesions. It is not practical where there are a very large number of lesions. A short burst of cryotherapy on the IGH lesions with a cryotherapy spray gun for about 5 seconds is sufficient. The repigmentation can take 6–8 weeks to develop. Ploysangam et al. [12] reported repigmentation in 90 % of treated lesions. Some lesions may develop hyperpigmentation after treatment but this improves with time. LN cryotherapy is fast, is very easy to perform as an office procedure and it is relatively less expensive.

Keywords

Cryotherapy • Idiopathic guttate hypomelanosis

Disease Description

Idiopathic guttate hypomelanosis (IGH) usually manifests as small, discrete, sharply demarcated, acquired hypopigmented or depigmented lesions measuring from 0.2 to 6 mm in diameter [1]. Rarely some lesions reach 20 mm [2]. The incidence of IGH increases with age and it is common in the elderly. Once IGH lesions appear spontaneous repigmentation does not seem to occur. Several clinical types of IGH have been described [1–10]. It affects all races. The commonest type is multiple hypopigmented or depigmented small discrete macules in the sun damaged skin of older individuals (Fig. 84.1).

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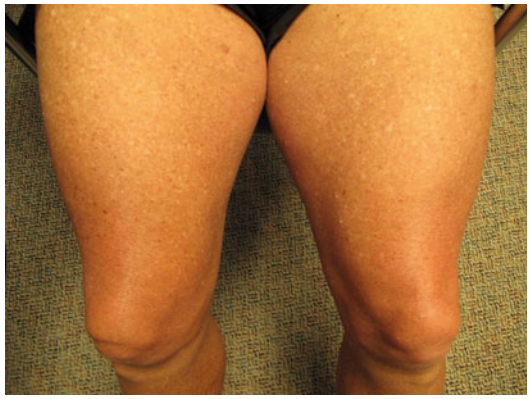

Fig. 84.1 An elderly woman with multiple hypopigmented IGH lesions on lower limbs on sun damaged skin

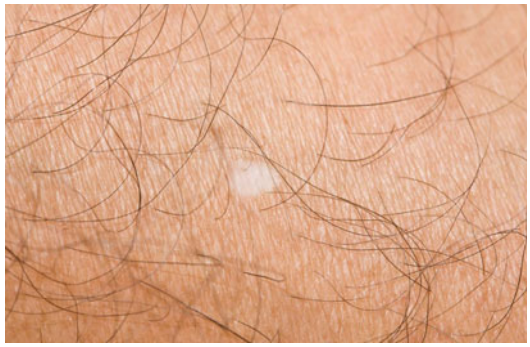

Fig. 84.2 Isolated porcelain white IGH lesion on the volar aspect of forearm in a person with Type IV skin

Some lesions may be porcelain white, whereas some are hypopigmented. A second type of IGH occurs as discrete lesions irrespective of sun damage; in exposed or non-sun exposed areas on the trunk or limbs (Fig. 84.2). This type is particularly noticeable in darker skinned individuals. Thirdly IGH can appear as small punctate (0.2–1 mm) lesions [8, 9]; sometimes following prolonged ultraviolet light therapy. Occasionally punctate IGH lesions are seen even without a history of UV light therapy. There is yet another type of IGH where there is a thin, macroscopically visible, pale keratinous layer over the hypopigmented lesions [1, 10]. When the keratinous layer is detached the typical appearance of IGH becomes visible [1]. Some of the IGH lesions appear sclerotic, particularly those that are porcelain white. IGH lesions rarely occur on the face [1] (Table 84.1). Chronic ultraviolet exposure due to sun exposure and aging are contributing factors at least in the common type of IGH on exposed areas.

Table 84.1 Types of IGH

Multiple hypopigmented or porcelain white lesions on sun-damaged skin
Solitary or multiple porcelain white lesions irrespective of areas of sun damage
Relatively smaller punctate leukoderma like lesions with or without a history of ultraviolet light treatment
Hypopigmented macules with a thin keratinous layer on top

Differential Diagnosis

Clinical differential diagnoses of IGH include: vitiligo, post-inflammatory hypopigmentation, pityriasis lichenoides chronica with secondary hypopigmentation, rain-drop depigmentation of chronic arsenic poisoning, dyschromic amyloidosis, extra-genital lichen sclerosus et atrophicus, pityriasis versicolor, depigmented macules in patients with tuberous sclerosus, dyschromatosis universalis, dyschromatosis symmetrica hereditaria, and hypopigmented macules in patients with Darier's disease. In most cases, clinical diagnosis of IGH is easy due to the characteristic appearance of the lesions. Wood's lamp examination may help identify the borders of a lesion. If in doubt a skin scrapings test for *Malassezia furfur* or a skin biopsy may be done.

Histopathology of IGH shows a reduction of melanisation of the keratinocytes in the area [4]. There is a significant reduction,or (rarely) a total absence of melanocytes in the lesions [4, 11, 12]. Within the lesions the melanin granules are irregularly and focally distributed [7]. Other histopathological features include basket weave hyperkeratosis, epithelial atrophy and flattening of the retepegs [4]. Occasionally there can be sclerotic changes in the dermal collagen. Electron microscopy of IGH lesions show round and less dendritic melanocytes with fewer melanosomes [11, 12]. Cell cultures of melanocytes from IGH lesions (on cell culture plates) are able to proliferate and produce melanin as normally

as non-lesional melanocytes. Some of the melanocytes show morphological abnormalities. Some senescence markers (e.g. p 21) are more expressed in the lesional melanocytes compared to non lesional melanocytes (Parsad D, Kumarasinghe SP, Kumar R, on going study). It is not yet clear whether the upregulation of senescence markers of IGH melanocytes is driven by other cells in the epidermal melanin unit milieu.

According to some studies, it is likely that in IGH there is an inhibitory influence on melanin transfer and/or production due to overlying keratinocytes [4, 8, 12]. Malfunctioning melanocytes may degenerate and be removed by the body. This theory is supported by the observation that when small punch grafts are transplanted in vitiligo patients the pigment spreads outwards, migrating even up to 5 mm around the graft, but IGH lesions with sharply demarcated borders resist pigment spread from surrounding normal melanocytes. Falabella et al. demonstrated that when a small piece of normal skin was grafted to an area of IGH (after removing a similar sized specimen from an IGH lesion) the whole punch graft eventually got depigmented rather than the IGH lesion getting pigmented from the grafted normal skin [4]. Support for this theory also comes from the fact that removal of the epidermis of IGH lesions by cryotherapy, dermabrasion or laser ablation, causes repigmentation (Fig. 84.3). It is not clear whether dermis of the IGH lesions is exerting any influence in the melanisation of the keratinocytes or destruction the melanocytes. The exact cause of loss of pigment in the IGH lesions is not yet known.

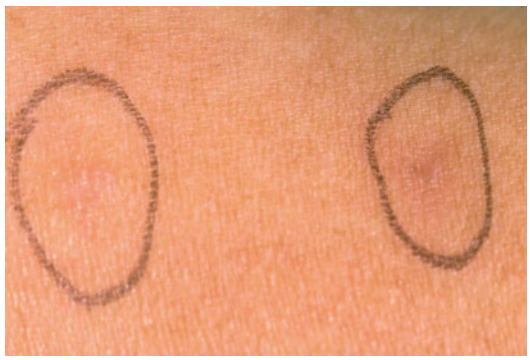

Fig. 84.3 A patient who had cryotherapy 6 weeks prior showing slight hyperigmentation of the lesion on the right, whereas the one on the left repigmented to the desired level

Cryotherapy

Ploysangam et al. initially described beneficial effects of cryotherapy in IGH [12]. They did cryotherapy for 15 s, using cryoprobes; all the patients developed vesicles. However, we showed in a case series that even 5-s short bursts of cryotherapy are adequate to bring the pigment back to the lesions [9]. The explanation for this would be that cryotherapy destroys the top layer of skin (epidermis). Cryotherapy also causes tissue inflammation in the lesional and perilesional skin, this too may have some stimulating effect on repigmentation. The short bursts of cryotherapy is better, particularly in the dark skinned patients due to possible leukoderma of the surrounding skin due to excessive cryotherapy. Furthermore it is less painful to the patients and causes only minimal or no vesicles. Cryotherapy is an effective low cost simple method of treatment. If required cryotherapy can be repeated after 6 weeks. However, when there are a large number of IGH lesions none of the treatment modalities are practically useful.

The mechanism of repigmentation following cryotherapy is not clear. Ploysangam et al. showed, in biopsy specimens from repigmented lesions 6 weeks after LN treatment, more melanin and more active dendritic melanocytes compared to pretreatment [9, 12]. However, the total number of melanocytes in the repigmented lesions remained lower than in normal skin [12]. It appears that the removal of the epidermis over the IGH lesion somehow facilitates the surrounding and lesional melanocytes to repigment the site. It is not clear whether tissue inflammatory cytokines released due to cryotherapy work synergistically in repigmentation. Considering that some melanocytes can get destroyed during cryotherapy, it appears that the effect of removal of the IGH epidermis is dominant in the facilitation of repigmentation.

Success Rates

Cryotherapy does not work in all cases of IGH. Also in some cases the pigmentation returns but slowly the same patches lose the pigment again. The hypopigmented lesions respond better than the porcelain white sclerotic lesions. Punctate lesions of <2 mm are difficult to treat with cryotherapy.

Treatment Options

Other methods of treatment for IGH include laser ablation using a carbon dioxide or Erbium laser, fractional carbon dioxide laser, intralesional triamcinolone injections, topical tacrolimus 0.1 % application, topical pimecrolimus application, topical retinoids and punch grafting with triamcinolone injection [1, 4, 9, 12–16].

Methodology (How I Do It)

1. Ask the patient to point out lesions that need to be treated (it is not practical to treat hundreds of lesions in one sitting)
2. Use a cryotherapy spray gun rather than cotton buds, preferably use the standard nozzle aperture sizes (e.g. 0.5–1mm). Local anesthesia is not required.
3. Directly spray for approximately 5 s keeping a distance of 0.5 cm to 1 cm between the nozzle and the skin of 1–1.5 cm. Cryocones are not required. The frozen lesion should become whitish due to ice crystal formation. Most treated lesions and the immediate surrounding rim of skin will become slightly erythematous, transiently. Usually, vesicles do not appear after treatment, however a few may occur. After a few days the top layer peels off.
4. During the initial few treatments a timer (e.g. 5 s) may be used, however it is not necessary for an experienced operator.
5. No dressings or specific care is needed. If vesicles occur, they are best left alone. Even if the vesicle roof comes off due to friction, the superficial erosions heal without any problems.
6. Repigmentation takes 6–8 weeks to fully develop. A new set of IGH lesions may be treated at each visit. If required, any lesions that have not repigmented adequately may be retreated.

Complications

Cryotherapy for IGH is a very simple procedure, usually without complications. The mild pain of freezing IGH lesions is well tolerated. Very rarely vesicles may become secondarily infected. Occasionally haemorrhagic vesicles can develop. Scarring does not occur unless frozen excessively. Rarely there can be secondary leukoderma in the surrounding skin. Some lesions may become hyperpigmented but this gets less pronounced with time.

Conclusions

Cryotherapy is effective in IGH. The procedure is simple, brief, easy to perform without anesthesia and relatively inexpensive. Some lesions may not respond to cryotherapy. Some may improve but later gradually lose pigment in the same area.

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