Dermatology Practical & Conceptual

Clinical Features of Vitiligo and Social Impact on Quality of Life

Julien Seneschal[1,2]

1 Department of Dermatology and Pediatric Dermatology, National Reference Center for Rare Skin disorders, Hôpital Saint-André, Bordeaux, France

2 University of Bordeaux, CNRS, Immuno ConcEpT, UMR 5164, Bordeaux, France

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Corresponding Author: Julien Seneschal M.D, Ph.D., Department of Dermatology, Hôpital Saint-André, CHU de Bordeaux, Bordeaux, France. Bordeaux University, CNRS, ImmunoConcept, UMR 5164, 33000 Bordeaux, France. Phone : +33(0)5 56 79 47 05 Fax : +33(0)5 56 79 49 75 E-mail : julien.seneschal@chu-bordeaux.fr

ABSTRACT Vitiligo is the most common cause of depigmentation and its estimated worldwide prevalence ranges from 0.5% to 2%. The disease is characterized by the development of white macules resulting from a loss of epidermal melanocytes. The term vitiligo (nonsegmental) is now a consensus umbrella term for all forms of generalized vitiligo. Two other subsets of vitiligo are segmental vitiligo and unclassified/undetermined vitiligo, which corresponds to focal disease and rare variants. A series of hypopigmented disorders may masquerade as vitiligo, and some of them need to be ruled out by specific procedures including a skin biopsy. The skin plays an important role in our interaction with the world and any change in the skin colour can have important psychological consequences. In this line, vitiligo has a major impact on quality of life.

In this review, we will detail the most recent data on the clinical features of vitiligo and its impact on quality of life.

Introduction

Vitiligo is the most common skin depigmenting disorder resulting from a selective loss of epidermal melanocytes and affects around 0.5-2 % of the world population. Both sexes are equally affected, and there are no apparent differences in rates of occurrence according to phototype or race [1,2]. Twenty five percent of cases are children with disease onset before the age of 10, the age of onset in paediatric series varies from 4 to 8 years. Very early onset, as young as 3 months, is acknowledged. The existence of true 'congenital vitiligo' remains controversial [3,4]. In fair-skinned individuals, vitiligo patches

are usually detected only after the first exposure of the skin to sunlight, following the first summer of life. The percentage of segmental vitiligo (SV) is higher in children compared to adults, whatever the ethnic background, suggesting a mosaic skin developmental predisposition. The prevalence of SV in childhood varies from 4.6 to 32.5% in published reports. In addition, besides the recognition of clinical lesions of vitiligo, it is now important to recognize clinical signs of disease that could justify for patients with a progressive disease a treatment to stop the spreading of the disease.

In addition, vitiligo is always associated with major impact on quality of life [5]. This review summarizes classification, clinical aspects, and current knowledge regarding the impact on quality of life of patients affected by vitiligo, a still high unmet disease.

Clinical Features of Vitiligo

Vitiligo is characterized by the progressive loss of melanocytes leaving white patches on the skin. It is usually diagnosed by clinical examination alone supported by Wood's lamp examination. The differential diagnoses of vitiligo include pityriasis alba, hypopigmented mycosis fungoides, tinea versicolor, idiopathic guttate hypomelanosis, and other hypo- or depigmented disorders. Additionally, the diagnosis of vitiligo could be difficult in patients with fair skin. In cases of uncertain diagnosis, a skin biopsy, mycologic examination, and appropriate blood tests may be needed to exclude a fungal infection, cutaneous lymphoma and other disorders. Routine screening of anti-thyroid antibodies and thyroid function are recommended. The term "vitiligo (non-segmental)" referring to all forms of vitiligo except segmental vitiligo is now proposed. Segmental vitiligo (SV) refers to a clinically unilateral segmental distribution of depigmented lesions. The coexistence of SV plus vitiligo (non-segmental) is called mixed vitiligo. Focal vitiligo, a term that applies to localized macules characterized by loss of melanocytes, was assigned to the category undetermined/unclassified until more definitive classification can be made on clinical grounds (generally after 1–2 years of follow-up).

Vitiligo Subsets

Generalized Vitiligo

This most common form of vitiligo is characterized by white macules involving multiple parts of the body, most often in a symmetrical pattern (Fig. 1). Skin hypopigmentation is usually asymptomatic, but a minority of patients mention preceding mild pruritus (probably due to the inflammation). The disease can start at any site of the body, but the fingers, hands, and face are frequently the initial sites. Depigmentation of scars is a common manifestation of the Koebner's phenomenon (mechanical induction of the disease, also by friction or chronic pressure by clothing or daily activities). Koebner's phenomenon is usually contemporary of disease flares [6]. Stable lesions are well demarcated. Mixed vitiligo is a more recently described, mostly paediatric subtype, with segmental involvement preceding typical generalized vitiligo [7]. The presence of leukotrichia and halo nevi have been noted as predictors of passage to mixed vitiligo in patients with SV [8]. Mixed vitiligo may exist in adults but is probably frequently masked by widespread bilateral lesions.

Acrofacial Vitiligo

In acrofacial vitiligo, the involved sites are usually limited to face, head, hands, and feet (Fig 1). A distinctive feature

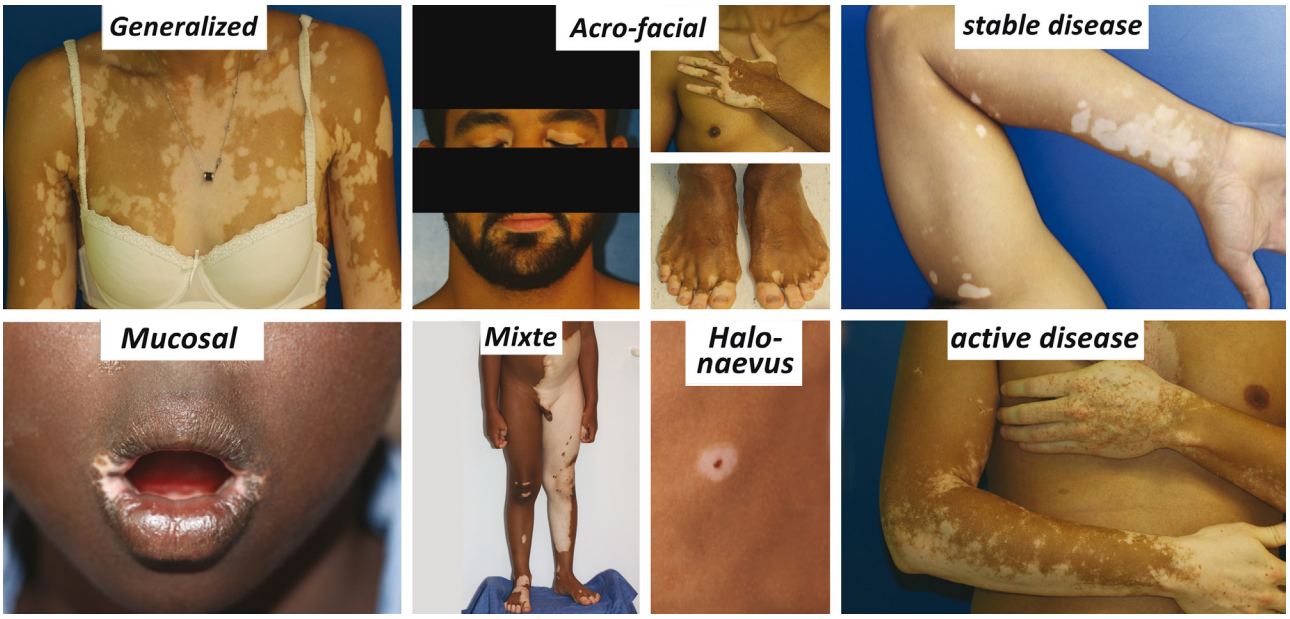

Figure 1. Clinical features of vitiligo

is depigmentation of the distal fingers and facial orifices. It may later include other body sites, resulting in typical generalized vitiligo. Acrofacial vitiligo was shown to be more frequent in adult onset cases of vitiligo in a large series studies using latent class analysis [9].

Vitiligo Universalis

Vitiligo universalis is a rare presentation of vitiligo. It is the most extensive form of the disease and generally occurs in adulthood. 'Universalis' is generally used when depigmentation is virtually universal (80–90% of body surface), but some pigmentation may be still present, and hairs partially spared.

Segmental Vitiligo

Mono-segmental vitiligo is the most common form of SV, referring to the presence of one or more white depigmented macules distributed on one side of the body, usually respecting the midline (although some lesions may partly cross the mid-line), early follicular involvement (leukotrichia), and rapid development over a few weeks or months, and overall protracted course but secondary extension remains possible in a given segment sometimes years after [10,11]. The aetiology of the SV pattern remains overall elusive. Rarely, multiple segmental lesions occur simultaneously or not distributed either unilaterally or bilaterally. A clear segmental distribution of the lesions with midline demarcation, together with the associated features described in mono-segmental cases (leukotrichia, protracted course), distinguishes this diagnosis versus vitiligo in bilateral cases.

Unclassified and Rare Variants

Focal cutaneous or mucosal vitiligo (defined as small isolated patch that does not fit a segmental distribution, and which has not evolved into vitiligo after a period of at least 2 years) should be left within the category undetermined/ unclassified vitiligo. Vitiligo ponctué/ punctata lesions present as sharply demarcated depigmented punctiform 1- to 1.5-mm macules involving any area of the body, and has to be distinguished histopathologically from guttate hypomelanosis, a common condition with no loss of melanocytes situated on chronically sun exposed sites such as the legs and forearms. Vitiligo minor/ hypochromic vitiligo to affect only dark-skinned individuals. 'Minor' refers to a partial defect in pigmentation. The relation to true vitiligo comes from pathology and coexistence with more typical vitiligo macules. Cutaneous T cell lymphoma needs to be ruled out by repeated biopsies with molecular studies of clonality, and this diagnosis cannot be made without a long-term follow-up. Follicular Vitiligo refers to a form of generalized vitiligo that primarily involved the pigment cell follicular reservoir with limited skin involvement, contrasting with marked generalized hair whitening.

Clinical Signs of Disease Activity

Recently it has been defined different clinical signs associated with the progression of the disease. Several visible clinical skin manifestations in vitiligo are reported in relation to disease progression such as inflammatory borders, Koebner phenomenon, hypochromic areas/ borders and confetti-like depigmentation [12]. The presence of these clinical markers in patients with vitiligo has been linked to poor prognosis, rapid disease progression and inadequate response to therapies. Confetti-like depigmentation was first described by Sosa JJ and defined as the presence of several grouped 1 to 5-mm depigmented macules, usually located at the border of an existing lesion [13]. Therefore, these signs are important to recognize to start as soon as possible treatment that could stop the spread of the disease (Fig. 2).

Social Impact on Quality of Life

Skin plays an important role in our interaction with the world, and skin colour is an important element in our interaction with the world. In this sense, any change in skin colour can have important psychological consequences. In patients with vitiligo, quality of life and disease burden can be measured by generic dermatology instruments such as the Daily Life Quality Index (DLQI). Although generic instruments such as the DLQI can provide a general picture of impaired quality of life, they generally fail to detect nuances in the way patients manage the overall burden of vitiligo. In this context, over the last decade, several specific scores to measure the individual impact of the disease have also been developed, such as the Vitiligo Impact Scale (VIS), the Vitiligo Quality Of Life (VitiQoL) or the Vitiligo Impact Patient Scale (VIPs) [14]. Of all these scores, only the VIPs considers the patient's phototype, which is a major factor in the experience of the disease [15]. In the late 1970s, Porter et al. first reported the major impact of vitiligo on patients' quality of life and there is now strong evidence that patients with vitiligo are negatively affected in their sexual relationships [16]. In addition, several studies have shown that adult patients with generalized vitiligo experience a decrease in quality of life comparable to that of patients with other skin diseases such as atopic eczema and psoriasis [5]. Indeed, many people are afraid and uncomfortable when they come into contact with people with vitiligo, thus discriminating against these patients. Vitiligo patients also complain that they do not receive enough support from their doctors. More recently, in 2005, a survey of members of the United Kingdom Vitiligo Society showed that over 50% of respondents said that vitiligo had a significant impact on their quality of life. In this study, finding an effective and long-lasting treatment was the top priority for the patients who responded to the survey and who had the most severe forms. Only a small number of

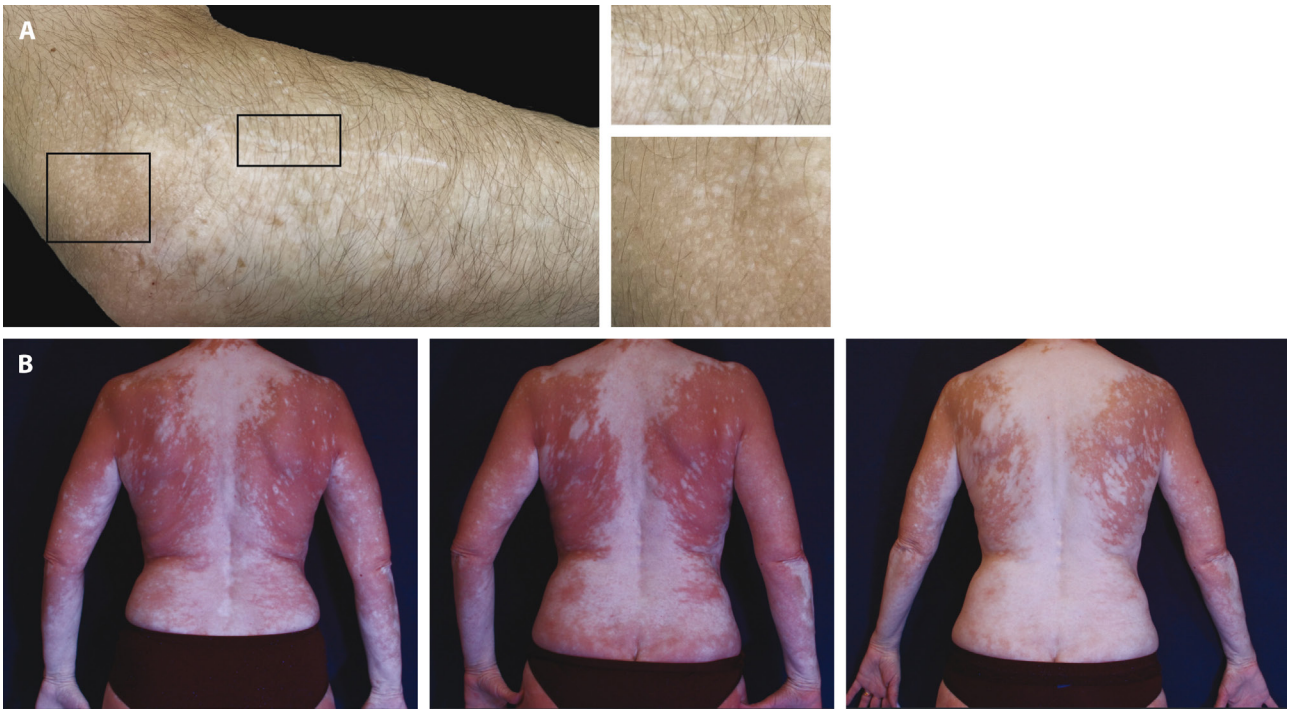

Figure 2. Clinical signs of disease activity. a. clinical images shows the presence of confettis-like lesions and koebner phenomenon type 2b (linear lesion on site of clinical friction). b. evolution of vitiligo without effective treatments in patients with active disease

respondents reported getting information about vitiligo from their dermatologist (12.5%), while more than 80% get this information from non-medical sources [17]. More generally, several factors can have an impact on the quality of life of vitiligo patients, namely age of onset, skin phototype, extent and distribution, impact of treatment, among others. For example, Nogueira et al. found that most vitiligo patients with affected areas corresponding to visible areas (88%) complain of negative emotions compared to those with lesions in non-visible areas (20%). Illness (71%), shame (57%), insecurity (55%), sadness (55%) and inhibition (53%) were the most frequently cited negative emotions in patients with visible lesions [18,19]. Similarly, Parsad et al. found that injuries to the face, arms, legs and hands had a greater impact on DLQI [20]. The degree of stigma is probably related to cultural background and may therefore explain some of the variation in DLQI observed between countries [21]. Specifically, the self-image of vitiligo patients is significantly impaired and mood insensitivity is common, especially in adolescents [22]. The psychological impact of vitiligo in childhood can be profound and have a lasting effect on personal self-esteem. Parsad et al. found that children with vitiligo were more likely to avoid/ restrict sports activities and miss more school days than those without vitiligo [20]. In another study from the Netherlands, it was shown that the impact of vitiligo occurring in childhood can persist into adult life with a social impact [23]. It is worth noting that a study on quality of life issues in childhood skin diseases found that people with vitiligo suffer from low self-esteem, social stigma, shame, avoidance of intimacy, anxiety, depression, adjustment disorders, fear, suicidal ideation and other psychiatric comorbidities [24]. In an internet survey of children aged 0- 17 years and their families, the deterioration in quality of life increased with age: more than 90% of teenagers (15-17 years) were bothered by vitiligo compared to 50% of children aged 0-14 years [22]. The localization of the disease vitiligo on the face, arms and legs was found to be the most distressing and was associated with teasing and bullying. Finally, depression and anxiety in children were reported by 26% and 42% of parents and caregivers respectively [25].

Conclusion

Vitiligo is a common disease with an estimated global prevalence of 1%. Its impact on quality of life and self-esteem is considerable and long-lasting, with possible repercussions in adulthood for children with the disease. In this context, it is important to listen to patients and to take charge of their psychological suffering by not hesitating to offer psychological care.

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