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Clinical Appearance of Vitiligo and Piebaldism: A Review Article

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ABSTRACT

Introduction: Vitiligo is idiopathic hypomelanosis characterized by the presence of white macules that can expand. Can affect all parts of the body that contain melanocyte cells, for example, hair and eyes. The cause is unknown, various trigger factors are often reported, for example emotional crisis and physical trauma. Pathogenesis of vitiligo still unclear, there is an association between vitilligo and Hashimoto's thyroditis, pernicious anemia, and hypoparathyroid melanocytes found in the serum of 80% of vitiligo patients. Moreover, there is disease that similar to vitiligo, called piebaldism. Patches of skin that do not contain pigment that is found at birth and persists for life.The disease is inherited autosomally, due to differentiation and possibly the melanoblast membrane.

Discussion: Inheritance of vitiligo can involve genes related to the biosynthesis of melanin, the response to oxidative stress and autoimmune regulation. The macula is white with a diameter of several millimeters to several centimeters, round or oval with defined boundaries, with no other epidermal changes. Hypomelanotic macules are sometimes seen in addition to apigmented macules. In vitiligo macules can be found macules with normal pigmentation or hyperpigmentation called perifollicular repigmentation. Occasionally there is a raised edge of the lesion, erythema and itching, which is called inflammatory. Piebaldism is In the form of skin patches that do not contain pigment on the forehead, median or paramedian, accompanied by white hair. White patches are sometimes also found on the upper chest, abdoment and legs. Normal skin color or hypermelanosis there are areas that are hypomelanosis. Ultrastructural investigation showed no visible melanocytes and melanosomes in hypomelanotic areas. On the other hand, hypemelanotic islets are found with melanocytes that produce melanosomes normally, but if abnormal spheric milanosomes and granules are found, abnormal spherical and ganular melanosomes are also found.

Conclusion: It is distinguished from vitiligo which usually does not appear at birth, the shape and distribution are also different. On piebaldism. In piebaldism accompanied by white forlock and the presence of islets with normal pigments in hypomelanotic areas. Differentiated from the nevus with depigmentosus, in the nevus the number of melanocit is normal. If piebaldism is accompanied by abnormalities in the distance of the two pupils or is accompanied by deafness, then the possibility of Waardenburg syndrome should be considered.

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Introduction

Vitiligo is idiopathic hypomelanosis characterized by the presence of white macules that can expand. Can affect all parts of the body that contain melanocyte cells, for example, hair and eyes. The cause is unknown, various trigger factors are often reported, for example emotional crisis and physical trauma. Pathogenesis of vitiligo still unclear, there is an association between vitilligo and Hashimoto's thyroditis, pernicious anemia, and hypoparathyroid melanocytes found in the serum of 80% of vitiligo patients [1].

Because melanocytes are formed from the neuralcrest, it is suspected that neural factors have an effect. Tyrosine is a substrate for the formation of melanin and catechols. It is possible that intermediate products formed during catechol synthesis have a detrimental effect on melanocytes. In some lesions there is disturbance of sweat and blood vessels to the response of nerve transmitters, eg acetylcholine [1,2].

Another risk factor is melanocyte cells form melanin by oxidation of tyrosine to DOPA and DOPA to dopakinone. Dopakinon will be oxidized into various indoles and free radicals. Melanocytes in vitiligo lesions are damaged by the buildup of the precursor melanin. In vitro it has been shown that tyrosine, dopa and dopaque are cytotoxic to melanocytes [3].

Skin depigmentation may occur from exposure to mono benzyl ether hydroquinone in gloves or detergents containing phenols. Oxidative stress also plays an important role in the pathogenesis of vitiligo. Some experts believe that the accumulation of free radicals is toxic to melanocytes which in turn can cause damage

to these melanocytes. In vitiligo patients and in vitro showed an increase in NO levels which led to autodestruction of melanocytes [4].

Moreover, there is disease that similar to vitiligo, called piebaldism. Patches of skin that do not contain pigment that is found at birth and persists for life.The disease is inherited autosomally, due to differentiation and possibly the melanoblast membrane [5].

Discussion

Clinical symptoms and Classification

Inheritance of vitiligo can involve genes related to the biosynthesis of melanin, the response to oxidative stress and autoimmune regulation. HLA may be associated with vitiligo and several studies have shown that several types of HLA are associated with vitiligo including A2, DR4, DR7, and Cw6 [6].

The macula is white with a diameter of several millimeters to several centimeters, round or oval with defined boundaries, with no other epidermal changes. Hypomelanotic macules are sometimes seen in addition to apigmented macules. In vitiligo macules can be found macules with normal pigmentation or hyperpigmentation called perifollicular repigmentation. Occasionally there is a raised edge of the lesion, erythema and itching, which is called inflammatory [7].

The areas often affected are the extensor bones, especially above the fingers, periorificials around the eyes, mouth and nose, tibialis anterior, and wrist flexor. Bilateral lesions may be symmetrical or asymmetrical. In traumatized areas, vitiligo may develop. The mucosa is rarely affected, sometimes affecting the external genitalia, nipples, lips and gingiva.
There are 2 forms of vitiligo [8,9]:

1. Localwhich can be further divided into:
a. Focal: one or more macules in one area, but not segmental
b. Segmental: one or more macules in one area, with a distribution according to the dermatome, for example one leg
c. Mucosal: found only in mucous membranes
2. General
   Nearly 90% of patients are generalized and usually symmetrical. Generalized vitiligo can be further divided into:
a. Acrofacial: depigmentation occurs only distal to the extremities and face, is the initial stage of generalized vitiligo.
b. Vulgaris: macula without a pattern in many places
c. Mixed: complete or almost complete depigmentation is total vitiligo

Diagnosis and Evaluation of Vitiligo

1. Clinical evaluation [10]
   The diagnosis of vitiligo is based on history and clinical features. History:
a. Onset of disease
b. Family history of early onset of lesions and gray hair
c. History of thyroid disorders, alopecia aerata, diabetes mellitus, and pernicious anemia
d. Possible precipitating factors, such as stress, emotions, sunburn, and chemical exposure
e. History of inflammation, irritation, or skin rash before the white patches

2. Histopathological examination

With hematoxylin eosin (HE) staining it appears normal unless there are no melanocytes, sometimes lymphocytes are found on the margins of the macula. The dopa reaction to melanocytes is negative in the apigmented areas, but increases at the hyperpigmented margins [1-5].

3. Biochemical examination

Histochemical examination of coolies incubated with dopa showed the absence of tyrosinase. Sometimes plasma and skin tyrosine is normal [6].

Management of Vitiligo

The differential diagnosis are piebaldism, Wardenburg syndrome, and woolf syndrome. Segmental vitiligo should be differentiated from nevus depigmentosus, tuberosclerosis, and hypomelanocytosis. Single or slight lesions should be differentiated from tinea versicolor, pitriasis alba, gutate hypomelanosis, and post-inflammatory hypopigmentation [9].

Vitiligo treatment is less than satisfactory. It is advisable for patients to use mask the disorder is covered with a cover mask. Systemic treatment is trimethylpsoralen or methoxy-psoralen in combination with sunlight or a light source containing long wave ultraviolet (ultraviolet A). The dose of psoralen is 0.6mg / kg BW 2 hours before exposure for 6 months to a year. Topical treatment with psoralen applied five minutes before exposure often results in irritant contact dermatitis. In some patients with high potency corticosteroids, for example betamethasone valerate 0.1% or clobetasol propionate 0.05% is effective in producing pigment [11,12].

Under 18 years of age, it is only treated topically with 1% lotion methoxalene diluted 1:10 with spirtus crushed. The liquid is applied to the lesions. After letting it sit for 15 minutes, then drying it in the sun for 10 minutes. Drying time is getting longer. What is desired is erythema, but not to show erosions, vesicles or bullae. At 18 years of age, if the skin disorder is generalized, the treatment is combined with metoxalene capsules (10 mg). The drug is taken 2 capsules (20 mg) 2 hours before drying, 3 times a week. If localized lesions are given only topical treatment. If after 6 months there is no improvement, the treatment is stopped and is considered a failure [13,14].

MBEH (monobenzylether of hydroquinone) 20% can be used for the treatment of vitiligo that covers an area of more than 50% of the skin surface and is not successful with psoralen treatment. If there is no contact dermatitis treatment is continued for up to 4 weeks for normal areas. Depigmentation can occur after 2-3 months and complete after 1 year. The possibility of returning to normal pigmentation in areas exposed to sunlight and in people with dark skin should be prevented with sunscreen [15].

Another way is surgery with skin grafts, both on the entire epidermis and dermis as well as only melanocyte cell cultures. The areas of the fingertips, lips, elbows and knees generally give poor treatment results. Tried to do repigmentation by means of tattoos with ferrous oxide in glycerol or alcohol [16].

Clinical symptoms and Examination of Piebaldism

In the form of skin patches that do not contain pigment on the forehead, median or paramedian, accompanied by white hair. White patches are sometimes also found on the upper chest, abdoment and legs. Normal skin color or hypermelanosis there are areas that are hypomelanosis [15].

Ultrastructural investigation showed no visible melanocytes and melanosomes in hypomelanotic areas. On the other hand, hypemelanotic islets are found with melanocytes that produce

melanosomes normally, but if abnormal spheric milanosomes and granules are found, abnormal spherical and ganular melanosomes are also found [16].

What is the difference With Vitiligo?

It is distinguished from vitiligo which usually does not appear at birth, the shape and distribution are also different. On piebaldism. In piebaldism accompanied by white forlock and the presence of islets with normal pigments in hypomelanotic areas. Differentiated from the nevus with depigmentosus, in the nevus the number of melanocit is normal. If piebaldism is accompanied by abnormalities in the distance of the two pupils or is accompanied by deafness, then the possibility of Waardenburg syndrome should be considered [17].

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